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Different characteristics of ventilator application between tracheostomy- and noninvasive positive pressure ventilation patients with amyotrophic lateral sclerosis

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Abstract

The aim of the study was to investigate the appropriate home ventilator settings for patients with amyotrophic lateral sclerosis (ALS).

In total, 71 patients with ALS, who had received either a noninvasive positive pressure ventilation (NIPPV) or tracheostomy positive pressure ventilation (TPPV), were included. Accordingly, patients were divided into 2 groups (the TPPV and NIPPV groups). We retrospectively evaluated the values used in home ventilators for patients with ALS, who had maintained a stable level of CO_2 on both the arterial blood gas analysis (ABGA) and transcutaneous blood gas monitoring. To measure the main outcome, we also investigated the actual body weight (ABW) and predicted body weight (PBW) of patients, and the following setting values of ventilators were also recorded: the inspired tidal volume (V_{Ti}), minute ventilation (MV), peak inspiratory pressure (PIP), positive end expiratory pressure (PEEP), and inspiratory time (T_{ins}).

V_{Ti} and MV showed a significantly positive correlation with both PBW and ABW of patients in the TPPV group. However, both V_{Ti} and MV had greater significant correlation with PBW than ABW in the TPPV group. In addition, V_{Ti} and MV did not show a significantly positive correlation with either PBW or ABW in the NIPPV group.

In patients with ALS, PBW was more useful for predicting V_{Ti} and MV than ABW. Moreover, it will be helpful to know the differences of setting values between TPPV and NIPPV, especially because ALS patients are usually treated with TPPV due to the initial difficulties associated with NIPPV.

Abbreviations: ABGA = arterial blood gas analysis, ABW = actual body weight, ALI/ARDS = acute lung injury/acute respiratory distress syndrome, ALS = amyotrophic lateral sclerosis, COPD = chronic obstructive pulmonary disease, CT = computed tomography, MRC = medical record council, MV = minute ventilation, NIPPV = noninvasive positive pressure ventilation, PBW = predicted body weight, PEEP = positive end expiratory pressure, PEG = percutaneous endoscopic gastrotomy, PIP = peak inspiratory pressure, T_{ins} = inspiratory time, TPPV = tracheostomy positive pressure ventilation, V_{Ti} = inspired tidal volume.

Keywords: amyotrophic lateral sclerosis, minute ventilation, tidal volume, ventilation

1. Introduction

Respiratory failure due to respiratory muscle dysfunction is a common problem in neuromuscular disease, and respiratory muscle weakness can be reversible, relapsing, or progressive.[1] Amyotrophic lateral sclerosis (ALS), which is a neurodegenerative disease characterized by progressive neuromuscular atrophy with early involvement of the respiratory system, may lead to pulmonary collapse that requires mechanical ventilation. It represents a major cause of mortality.[2]

Noninvasive positive pressure ventilation (NIPPV) or tracheostomy positive pressure ventilation (TPPV) is usually used to treat neuromuscular disease with symptomatic hypoventilation.[3,4] Among these 2 methods, NIPPV has been widely recommended for patients with neuromuscular disease accompanied by chronic respiratory failure, since it not only reduces dyspnea and improves persistent hypoventilation, but it may also extend the life of individuals affected by this fatal disease.[4] Conversely, TPPV is used to treat ALS patients with severe bulbar palsy unable to maintain a proper level of CO_2 via NIPPV.[5]

Among the parameters of mechanical ventilation, the tidal volume is one of the more important parameters. Over the past decades, the tidal volume of invasive ventilation has progressively decreased, from greater than 12 to 15 mL/kg to less than 9 mL/kg of the actual body weight in patients with acute lung injury/acute respiratory distress syndrome (ALI/ARDS).[6,7] Currently, there are guidelines that strongly support the use of a lower tidal volume (i.e., 6 mL/kg predicted body weight) in patients with ALI/ARDS.[8] However, there is no consensus on the optimal initial tidal volume for patients without ALI/ARDS, especially in ALS.[9]

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Moreover, a lower tidal volume may cause atelectasis due to insufficient ventilation in patients with respiratory muscle weakness, such as ALS. As a result, clinicians who are not experts on ventilator settings may experience difficulty in using ventilators, which may lead to inappropriate application.

Therefore, in efforts to determine the appropriate ventilator settings, we retrospectively evaluated the values used in home ventilators for patients with ALS, and had a stable level of CO_2 on both the arterial blood gas analysis (ABGA) and transcutaneous blood gas monitoring (Sentec AG, Therwil, Switzerland).

2. Materials and methods

This retrospective, single-center study was approved by the appropriate ethics committee. A total of 71 patients with ALS and respiratory failure, who were ventilator dependent at home and at the rehabilitation department, between June 2010 and December 2015, were included for evaluation. To reduce bias due to the ventilator mode, patients with ALS receiving a volume control mode (AC mode) with home ventilator Triology 100 (Philips Respironics, Murrysville, PA) was investigated in our study. Home ventilators were applied to all patients using an unvented circuit. Home ventilators were applied to patients at our clinic when they exhibited symptoms of respiratory insufficiency and hypercapnia, defined as partial pressure of arterial carbon dioxide ($PaCO_2$ and $PtCO_2$) > 45 mm Hg via arterial blood gas analysis (ABGA) and transcutaneous blood gas ($PtCO_2$) analysis; SenTec Digital Monitor System with V-Sign Sensor was used for the latter (Sentec AG, Therwil, Switzerland).[11,12] Patients were routinely admitted when they needed to control the ventilator setting, and ABGA and $PtCO_2$ monitoring was performed during admission. The ABGA was performed routinely at 6 AM to evaluate night time hypercapnia. Moreover, $PtCO_2$ monitoring was performed continuously for more than 48 hours before and after the application of the ventilator. In our clinic, NIPPV via a nasal mask was applied initially to patients with hypercapnia. If the nasal mask was insufficient, they switched to a facial mask to improve respiratory insufficiency and hypercapnia. In addition, TPPV was applied when respiratory insufficiency and hypercapnia were not improved by NIPPV. In TPPV, a cuffed tracheostomy tube with 10 to 20 mm Hg of cuff pressure was used in our clinic.

We retrospectively reviewed the charts of patients to investigate the appropriate ventilator parameters for patients with ALS. The following parameters of ventilators were successfully collected: ventilator mode, inspired tidal volume (V_{Ti}), actual minute ventilation (MV), peak inspiratory pressure (PIP), positive end expiratory pressure (PEEP), and inspiratory time (T_{ins}). The presence of sustained clinical improvement with normal pH (≥7.35 and ≤7.45), $PtCO_2$ and $PaCO_2$ (≤45 mm Hg and ≥35 mm Hg), and oxygen saturation (≥ 90%), in addition to the lack of atelectasis in chest imaging (chest x-ray and CT) were required to be considered as successful TIPPV or NIPPV.[4,13] In addition, the following parameters were also collected for every patient: age, actual body weight (ABW), predicted body weight (PBW) calculated using height (Male; [height(cm) – 152.4] × 0.91 + 50, female; [height (cm) – 152.4] × 0.91 + 45.5), presence of tracheal tube and percutaneous endoscopic gastrotomy (PEG), and duration of ventilator usage per day (in hours). Moreover, the duration of disease and Medical record council (MRC) scales of the upper and lower extremities were also checked to investigate the relevance associated with the course of the disease.[14] To assess the functional status of patients at presentation such as ambulatory function and the degree of disability, the modified Rankin scale was also checked.[15]

The exclusion criteria were as follows: (1) patients with ventilators operating in pressure cycled mode; (2) patients with sustained respiratory failure symptoms, such as headache, dyspnea, day somnolence with abnormal pH (<7.35), $PtCO_2$ or $PaCO_2$ (>45 mm Hg) and oxygen saturation (<90%), despite the use of a ventilator; (3) patients with massive retention of secretion, pneumonia, or other infection, such as urinary tract infection; (4) patients with hemodynamic instability; (5) patients with coma or impairment of consciousness (Kelly scale >2)[16]; (6) patients with atelectasis as shown on chest imaging (chest x-ray and CT); and (7) patients with chronic lung disease, such as chronic obstructive pulmonary disease (COPD), emphysema, and old tuberculosis.

Of the total 71 study participants, 29 patients were excluded; 16 patients for incomplete medical records (such as missing several ventilator parameters or body weight), another 10 patients for receiving ventilators in pressure control mode, and the remaining 3 patients for the detection of atelectasis on chest computed tomography (CT) (Fig. 1). The remaining patients were divided retrospectively into 2 groups—the first group used TPPV with a tracheal tube as the treatment method (the TPPV group), and the second group used NIPPV with a facial mask as the treatment method (the NIPPV group). The demographic data for all patients are summarized in Table 1.

2.1. Statistical analysis

We compared the characteristics between the 2 groups using Mann Whitney *U*-tests, where appropriate. Pearson correlations analysis was used to find the variables with significant correlations between the ventilator settings and patient's factors. To develop ventilator equations, a univariate linear regression analysis was used. V_{Ti} and MV were considered as dependent variables, and ABW and PBW, which had the highest Pearson correlation coefficient with V_{Ti} and MV, were considered as independent variables. In addition, to calculate the statistical significance of the difference between R^2 values of each regression model, each regression was repeated using bootstrap resampling with 1000 iterations. Then, the Wilcoxon signed rank test was performed for the *P*-value between the 2 groups (ABW- vs PBW-estimate). Using the same method, we also calculated the statistical significance of the difference between R^2 values in regression models with and without a constant.

Statistical analyses were performed using R software 3.1.1. and SPSS for Windows[c] (version 2.15.2, R Foundation for Statistical Computing, Vienna, Austria).

3. Results

3.1. Demographic and setting values of ventilator

Table 1 shows the demographic and ventilator settings. There were significant differences in the duration of ventilation, V_{Ti}, MV, PEEP, MRC scales of upper and lower extremities, and duration of disease between the TPPV and NIPPV groups. A modified Rankin scale in the NIPPV group was significantly lower than that in the TPPV group. V_{Ti} in NIPPV was about 214 mL higher than V_{Ti} in TPPV. Moreover, MV in the NIPPV group was also higher by about 3.3 L/min than that in the TPPV group (Table 1).

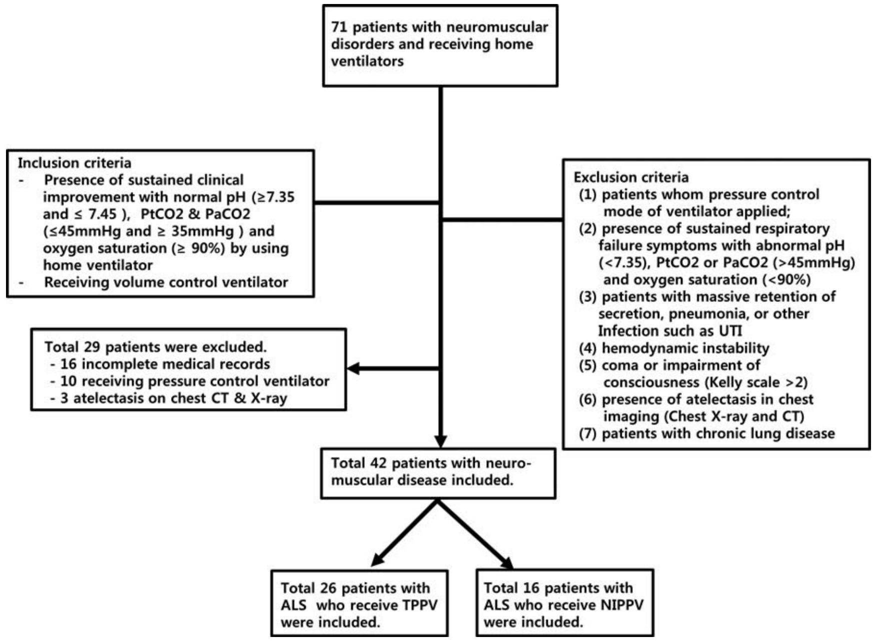

Figure 1. Flowchart of this study.

Regarding PIP, however, there was no significant difference between the NIPPV and TPPV groups. In the volume cycled ventilator, both NIPPV and TPPV groups had a similar value of PIP (14.34±4.9 vs 16.55±4.29).

Table 1

Demographic and parameters of home ventilator in both of TPPV and NIPPV groups.

Characteristics	TIPPV	NIPPV
Number, male: female	26 (17:9)	16 (10:6)
Age, y	69.12±11.84	69.81±10.99
Diagnosis, number	Limb onset ALS (13)	Bulbar onset ALS (8)
	Bulbar onset ALS (13)	Limb onset ALS (8)
Parameters of patients		
Height, cm	163.80±10.75	162.73±5.03
Body weight, kg	53.06±11.70	53.48±13.55
Duration of disease, months	59.77±42.71	26.94±17.95*
Usage time of ventilator, h/d	24.00±0.00	12.63±7.11*
MRC scale of shoulder abduction	1.68±1.35	2.75±1.57*
MRC scale of elbow flexion	1.68±1.35	2.94±1.57*
MRC scale of finger flexion	1.68±1.31	2.94±1.56*
MRC scale of hip flexion	1.96±1.33	3.06±1.29*
MRC scale of knee extension	1.96±1.40	3.13±1.31*
MRC scale of ankle DF	1.84±1.31	2.81±1.42*
Modified Rankin Scale	4.44±0.71	3.69±0.94*
Parameters of ventilator		
Inspired tidal volume, mL	485.5±124.17	699.37±178.41*
Peak inspiratory pressure, mm Hg	16.55±4.29	14.44±4.76
Minute ventilation, L/min	7.40±2.12	10.72±2.80*
Respiratory rate, number/min	15.08±2.54	16.46±2.94
Inspiration time, s	1.30±0.22	1.21±0.11
PEEP, mm Hg	4.36±0.91	2.55±2.50*

Values are mean±SD or as otherwise indicated.
ALS=amyotrophic lateral sclerosis, DF =dorsi-flexion MRC=medial research council, NIPPV= noninvasive positive pressure ventilation, PEEP=positive end-expiratory pressure, SD = standard deviation, TIPPV=tracheal intermittent positive pressure ventilation.
* <0.05 a significant difference compared to the TIPPV group.

3.2. Development of the equations for estimating V_{Ti} and MV

In a regression model, we selected independent variables based on the results from correlations analyses. Out of the 11 parameters, ABW had significant correlations with V_{Ti} and MV in the NIPPV group. However, PBW had a significant correlation with only V_{Ti} in the NIPPV group. In the TPPV group, both ABW and PBW had a higher correlation with V_{Ti} and MV compared with other parameters (Table 2). As a result, independent variables included ABW and PBW.

In the NIPPV group, V_{Ti} and MV were calculated by a univariate regression analysis between the ABW and PBW. However, V_{Ti} and MV had no significantly positive correlations with both PBW and ABW in the NIPPV group ($P \geq 0.05$) (Table 3) (Fig. 2).

In the TPPV group, V_{Ti} and MV were calculated by a univariate linear regression analysis between the ABW and PBW. In the TPPV group, V_{Ti} and MV had significantly positive correlations with both PBW and ABW ($P<0.05$) (Table 3) (Fig. 3).

3.3. Comparison of ABW-estimated and PBW-estimated regression models

In the regression models with a constant, PBW had a greater correlation with V_{Ti} and MV than ABW in the TPPV group (adjusted $R^2=0.678$ vs 0.268 in V_T, adjusted $R^2=0.513$ vs 0.274 in MV) (Table 4). In the result of Wilcoxon signed rank test, which was performed for the P-value between R^2 values of 2 regression models (ABW-estimated vs PBW-estimated), the PBW-estimated regression models and MV had significantly larger R^2 values than ABW-estimated regression models ($P<0.001$ in TV, and $P<0.001$ in MV) (Table 4). In the regression models without a constant, the PBW-estimated regression models and MV had significantly larger R^2 values than ABW-estimated regression models ($P<0.001$ in TV_i, and $P<0.001$ in MV) (Table 4).

Table 2

Correlation coefficients between parameters of patients and setting values of ventilator in both NIPPV and TPPV groups.

	NIPPV group		TPPV group	
	Tidal volume	Minute ventilation	Tidal volume	Minute ventilation
Predicted body weight, kg	0.536*	0.255	0.783†	0.675†
Actual body weight, kg	0.557*	0.559*	0.649†	0.574*

Values are mean ± SD or as otherwise indicated.
NIPPV = noninvasive positive pressure ventilation, SD = standard deviation, TIPPV = tracheal intermittent positive pressure ventilation.
* $P<0.05$.
† $P<0.0001$.

3.4. Comparison of regression models with and without a constant

In the result of Wilcoxon's signed rank test, which was performed for the P-value between the R^2 values of regression models with and without a constant, the R^2 values of regression models without a constant were significantly larger than the regression models with a constant ($P<0.001$ in the PBW-estimated regression model for V_T, and $P<0.001$ in the PBW-estimated regression model for MV).

Table 3

Linear regression models for tidal volume and minute ventilation.

	Tidal volume (95% CI)	Minute ventilation (95% CI)
TPPV group		
PBW	9.466 (6.801 to 12.131)†	0.142 (0.086 to 0.198)†
ABW	5.791 (2.042 to 9.540)*	0.100 (0.036 to 0.163)*
NIPPV group		
PBW	12.867 (−1.558 to 27.292)	0.107 (−0.140 to 0.353)
ABW	6.009 (−0.708 to 12.726)	0.091 (−0.015 to 0.197)

The linear regresion model to estimate the effect of PBW and ABW on tidal volume and minute ventilation.
The presented variables are estimated B with 95% confidence interval.
ABW = actual body weight, CI = confidence interval, NIPPV = noninvasive positive pressure ventilation, PBW = predicted body weight, TPPV = tracheostomy positive pressure ventilation.
* $P<0.05$.
† $P<0.001$.

3.5. The changes of ventilator setting when NIPPV was changed to TPPV

We added the changes of MV, V_{Ti}, and PIP in 5 patients with ALS when NIPPV was converted to TPPV (Table 5). On average, V_{Ti} in NIPPV was about 240 mL higher than that in TPPV. However, the PIP remained similar when NIPPV was changed to TPPV

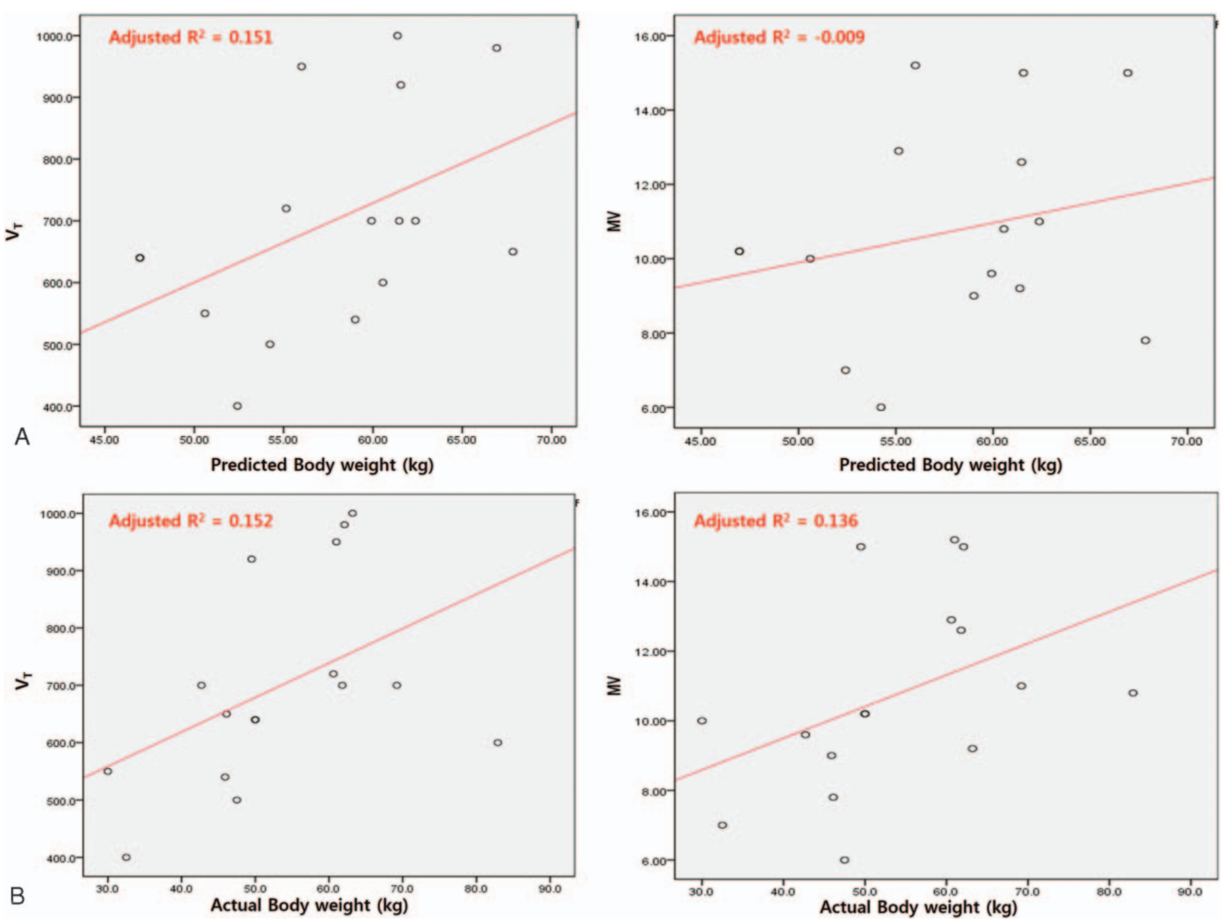

Figure 2. Estimation of tidal volume and minute ventilation in the NIPPV group. (A) Estimation of tidal volume and minute ventilation by using the predicted body weight. (B) Estimation of tidal volume and minute ventilation by using the actual body weight. Red solid line: regression line. MV = minute ventilation, NIPPV = noninvasive positive pressure ventilation, V_T = tidal volume.

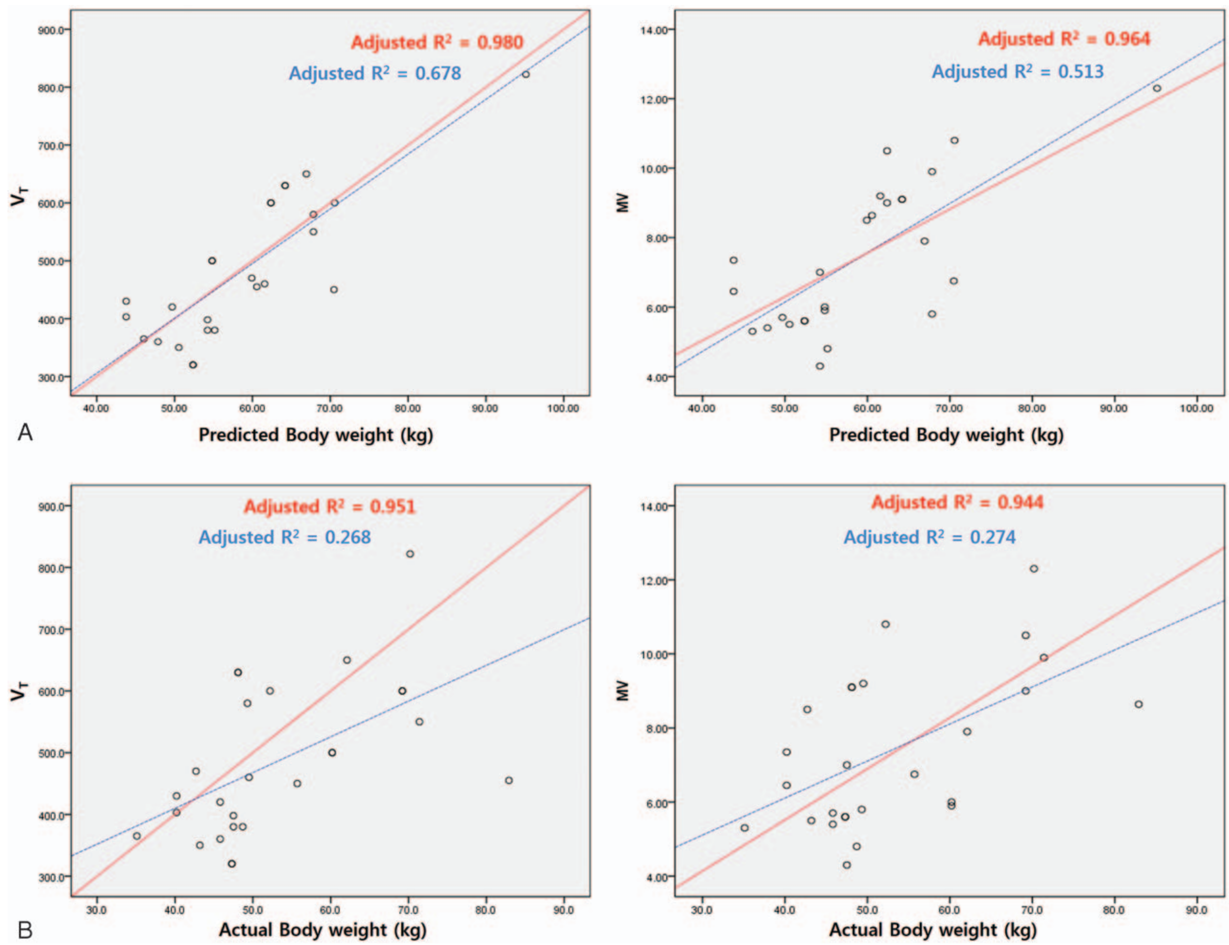

Figure 3. Estimation of tidal volume and minute ventilation (MV) in the TPPV group. (A) Estimation of tidal volume and minute ventilation by using the predicted body weight. (B) Estimation of tidal volume and minute ventilation by using the actual body weight. Red solid line: regression line without a constant. Blue dotted line: regression line with a constant. MV = minute ventilation, TIPPV=tracheal intermittent positive pressure ventilation, V_T = tidal volume.

(Table 5). In addition, there was no significant difference in the PIP between the NIPPV and TPPV groups (NIPPV, 16.55±4.29 (minimum 8.8– maximum 25.0); TPPV, 14.44±4.76 (minimum 10.0–maximum 27.4).

4. Discussion

Although several studies have developed equations to estimate the tidal volume, most of these studies were conducted based on subjects without neuromuscular diseases,[17,18] and there were

Table 4

Parameters estimates for regression models predicting (A) tidal volume and (B) minute ventilation in the TPPV group.

Regression models with a constant					
Dependent variables	**Parameter estimate**	**Bootstrap parameter estimate (95% CI)**	**Adjusted R^2**	**Bootstrap adjusted R^2 (95% CI)**	**P^***
(A) V_T					
PBW	9.466	9.671 (9.584–9.759)	0.678	0.664 (0.657–0.672)	<0.001
ABW	5.791	6.291 (6.143–6.440)	0.268	0.305 (0.296–0.315)	
(B) MV					
PBW	0.142	0.145 (0.143–0.146)	0.513	0.508 (0.499–0.517)	<0.001
ABW	0.100	0.102 (0.100–0.104)	0.274	0.289 (0.281–0.297)	
Regression models without a constant					
(A) V_T					
PBW	8.270	8.277 (8.263–8.290)	0.980	0.981 (0.980–0.981)	<0.001
ABW	9.000	9.035 (9.007–9.064)	0.951	0.954 (0.953–0.955)	
(B) MV					
PBW	0.126	0.126 (0.126–0.126)	0.964	0.965 (0.965–0.966)	<0.001
ABW	0.138	0.138 (0.138–0.138)	0.944	0.947 (0.946–0.947)	

* Wilcoxon signed rank test was performed between R^2 values between ABW- and PBW-estimated regression models.

ABW=actual body weight, CI = confidence interval, MV=minute ventilation, PBW=predicted body weight, TPPV=tracheostomy positive pressure ventilation, V_T=tidal volume.

Table 5

The changes of V_T and MV when NIPPV was changed to TPPV in patients with neuromuscular disease.

		NIPPV					TPPV				
	Disease	Date	mRS	Tidal volume (mL)	Minute ventilation (L/min)	PIP	Date	mRS	Tidal volume (mL)	Minute ventilation (L/min)	PIP
Patient 1	Limb onset ALS	2010–02–11	3	980	15	16	2015–08–11	5	650	7.9	14.3
Patient 2	Bulbar onset ALS	2014–11–26	2	700	9.6	15	2015–08–28	3	470	8.5	15.4
Patient 3	Bulbar onset ALS	2010–03–10	2	600	10.8	18	2010–07–14	3	455	8.6	18
Patient 4	Limb onset ALS	2014–12–03	2	500	6	12	2015–08–13	5	390	4.3	13
Patient 5	Limb onset ALS	2013–09–04	3	800	11	18	2015–08–26	5	600	9	18

ALS = amyotrophic lateral sclerosis, mRS = modified Ranke scale, MV = minute ventilation, NIPPV = noninvasive positive pressure ventilation, PIP = peak inspiratory pressure, TPPV = tracheostomy positive pressure ventilation, V_T = tidal volume.

only a few studies investigating the appropriate ventilator parameters for patients with amyotrophic lateral sclerosis. Unlike other neuromuscular diseases, ALS patients with pronounced bulbar symptoms often experience difficulty with NIPPV. Moreover, patients with ALS tend to have a faster disease progression. As disease progresses, it is not uncommon to change the ventilator application from NIPPV to TPPV in patients with ALS. Therefore, it may be unreasonable for patients with ALS to blindly undergo protocols and ventilator settings of other neuromuscular diseases uniformly, despite the existence of some protocols that outline the initial ventilator settings for neuromuscular disease.[19]

In this study, the equations using PBW for the calculation of an appropriate tidal volume were more accurate than ABW. This finding is consistent with previous studies demonstrating that the tidal volume based on PBW is more useful in predicting tidal volume than ABW.[20]

In this study, the categorized MV as well as V_T were calculated. Among the regression models used in this study, the R^2 value of the equation using PBW for calculating an appropriate tidal volume "estimated V_T (mL) = 8.27 × PBW (kg)" was the highest, followed by the R^2 value of the equation using PBW for calculating an appropriate minute ventilation "estimated MV (L/min) = 0.126 × PBW (kg)." Home ventilators with unvented circuit (Trilogy100; Philips Respironics, Murrysville, PA) used in our hospital, however, do not monitor the expired tidal volume (V_{Te}); they measure only V_{Ti}.[21] Thus, if V_T is adjusted only by considering PIP, it is possible for a clinician, who is not familiar with home ventilators, to think that the ventilator is malfunctioning. Such a misunderstanding may arise since low PIP, despite sufficient V_{Ti} could be developed in the following 2 cases: (1) patients with a lot of leakage volume and (2) patients with relatively sufficient respiratory muscle strength. Therefore, it is important to check not only PIP and V_{Ti}, but also V_{Te} or MV. As such, in the application process of home ventilators, it may be helpful if a clinician knew in advance of the appropriate tidal volume and MV using these equations, especially in patients with TPPV. Moreover, MV might be more useful in the initial settings regardless of the disease course, simply because MV tends to remain relatively constant, unlike other parameters, that is, worsening respiratory muscle weakness, decreased spontaneous breathing, and increased duration of ventilator usage, which are associated with neuromuscular disease progression.[10]

In this study, V_{Ti} and MV showed no statistically significant correlations with both PBW and ABW in the NIPPV group. This result shows that there may be a significant amount of air leakage through the facial mask. Since the leakage volume varies widely depending on the facial structures or the severity of the bulbar palsy, it is very difficult to quantify the leakage volume. Interestingly, however, PIP in 5 ALS patients remained similar even after changing the ventilator application from NIPPV to TPPV (Table 4). In addition, there was no significant difference in PIP between the NIPPV and TPPV groups (NIPPV, 16.55 ± 4.29 (minimum 8.8–maximum 25.0); TPPV, 14.44 ± 4.76 (minimum 10.0–maximum 27.4). These results suggest that PIP can be a relatively constant and important parameter when compared with V_T regardless of the severity of bulbar palsy during the initial application of NIPPV or when changing the ventilator from NIPPV to TPPV in patients with ALS.

Our present study had some limitations. First, only a small number of patients with various stages of ALS were included for evaluation. The leakage volume, which is affected by the degree of bulbar palsy, may have influenced our results. However, we tried to reflect the degree of disability by using a modified Ranke scale and by grouping patients based on the the ventilator application methods. Moreover, ALS is relatively rare, and encountering patients with such a disease and receiving ventilator treatment are even rarer. Considering the scarcity of neuromuscular diseases, this study might still be meaningful despite the small number of patients included for evaluation. Further studies that incorporate a greater number of patients with the same disease may be necessary. To analyze the effective usefulness of the categorized MV or V_T in the initial setting of home ventilator, a future prospective study may be necessary. Second, we only included TPPV and NIPPV with facial masks, not including those with mouthpieces and nasal masks. We were unable to quantify the leakage volume due to the retrospective nature of this study, especially in the NIPPV group. Therefore, we did not include NIPPV with mouthpieces or nasal masks to minimize the variable of leakage volume. In NIPPV, nasal masks are not recommended in an acute setting because it tends to lead to mask failure in >72% of subjects and seems to lower CO_2 to a lesser extent compared with facial masks.[22–24] In the case of mouthpieces, despite its comfort, there is a greater risk of hypoventilation.[25] Further studies that include other various types of ventilators might be helpful to better compare the effectiveness of each type. Lastly, we only included 5 ALS patients who underwent a change in the ventilator application from NIPPV to TPPV. Although 5 ALS patients showed interesting results with relatively constant PIP, even after changing the ventilator application, we were unable to generalize this finding in patients with ALS due to this limitation. However, the findings of this study showed that PIP may be an important parameter in ventilator settings regardless of the leakage, especially when NIPPV is applied to patients with ALS. In addition, it will be helpful to know the differences of setting values between TPPV and NIPPV, especially because ALS patients are usually treated with TPPV due to the initial difficulties associated with NIPPV. Further long-term clinical trials with larger numbers of patients are required to confirm these findings in this study.

5. Conclusions

In patients with ALS, appropriate management planning, including the use of TPPV or NIPPV, may reduce morbidity and mortality, and prevent the loss of lung compliance and atelectasis. In this study, equations using PBW, compared with those using ABW, were shown to be more accurate. This finding is consistent with previous studies. In NIPPV, V_{Ti} and MV had no significantly positive correlations with both PBW and ABW. However, PIP can be a relatively constant and important parameter compared with V_T, regardless of the severity of bulbar palsy during the initial application of NIPPV or when changing the ventilator application from NIPPV to TPPV in patients with ALS; a further prospective study may be necessary to confirm such findings. Moreover, it will be helpful to understand the differences of setting values between TPPV and NIPPV, especially because ALS patients are usually treated with TPPV due to the initial difficulties associated with NIPPV.

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