Respiratory Muscle Strength and Gas Exchange in Neuromuscular Diseases: Comparison with Chronic Pulmonary Emphysema and Idiopathic Pulmonary Fibrosis

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NISHIMURA, Y., HIDA, W., TAGUCHI, O., SAKURAI, M., ICHINoSE, M., INoue, H. and TAKISHIMA, T. Respiratory Muscle Strength and Gas Exchange in Neuromuscular Diseases: Comparison with Chronic Pulmonary Emphysema and Idiopathic Pulmonary Fibrosis. Tohoku J. Exp. Med., 1989, 159 (1), 57-68 — To examine whether or not the respiratory muscle weakness is correlated with decrease in arterial oxygen tension (PaO₂), respiratory muscle and pulmonary functions in 14 patients with neuromuscular diseases (NMD) were studied and compared with those of 12 patients with chronic pulmonary emphysema (CPE) and 15 patients with idiopathic pulmonary fibrosis (IPF). Respiratory muscle strength was assessed by maximal static inspiratory and expiratory mouth pressure at three lung volumes (RV, FRC and TLC). Although mean pulmonary functions in NMD showed virtually normal function, respiratory muscle strength was significantly less than the corresponding values in CPE and IPF. In NMD, maximal inspiratory mouth pressure at RV level (PImax) correlated positively with %TLC and %VC (r=0.652 and r=0.536, respectively). Moreover, PImax was significantly correlated with PaO₂ (r=0.561), but not with PaCO₂. Maximal expiratory mouth pressure at TLC (PEmax) correlated positively with %TLC and %VC. In CPE and IPF, respiratory muscle strength had no correlation with PaO₂ and PaCO₂. These findings suggest that inspiratory muscle dysfunction in NMD may be one of the factors responsible for determination of the level of hypoxemia and lung volume. ——— respiratory failure; pulmonary function test; maximal mouth pressure; hypoxemia and lung volume

It has been reported that patients with neuromuscular diseases (NMD) exhibit hypoxemia without hypercapnia in the early stage (Campbell 1965; Gibson et al. 1977), and both hypoxemia and hypercapnia in the advanced stage (Braun et al. 1977).
1983). These changes in blood gas exchange may be due to respiratory muscle weakness. Braun et al. (1983) found that respiratory muscle strength was significantly correlated with the degree of hypercapnia. However, the relationship between respiratory muscle strength and hypoxemia remains unknown.

In the present study, we examined the relationship between respiratory muscle functions and blood gases in NMD and compared the results with those from chronic pulmonary emphysema (CPE) and idiopathic pulmonary fibrosis (IPF). We found that maximal inspiratory muscle strength at residual volume (RV) was correlated with the degree of hypoxemia but was not with hypercapnia. This may support the conjecture that inspiratory muscle dysfunction causes hypoxemia.

**METHODS**

*Patient population*

We studied 14 patients with NMD (9 males and 5 females, mean age ±s.d., 52.6±11.1 year), 12 patients with CPE who were diagnosed by evaluation of alveolar destruction from selective alveolo-bronchograms (Aoki et al. 1984) (11 males and one female, 62.3±7.8 year) and 15 patients with idiopathic pulmonary fibrosis (IPF) (8 males and 7 females, 48.5±11.2 year). In the NMD, there were seven patients with amyotrophic lateral sclerosis, two with myasthenia gravis, four with myotonic myodystrophy, one with progressive myodystrophy and one with undetermined motor neuron disease. Each patient was in a stable state and free of acute exacerbation, although seven of the NMD patients, all of the CPE patients and ten of the IPF patients complained of exertional dyspnea. Informed consent was obtained from each subject for this protocol.

*Respiratory muscle function test*

Maximal static inspiratory and expiratory mouth pressure was measured in all patients according to the technique of Black and Hyatt (1969) using a device developed by us (VITALOPOWER KH101, Chest, Tokyo). This device consists of two parts; a plastic cylinder and a calculator. The cylinder has a closed end with a strain gauge pressure sensor and a small side hole which minimizes oral pressure artifacts. The opposite end is fitted with a mouthpiece. The time-pressure curve is displayed on a chart. Maximal inspiratory mouth pressure was measured at levels of RV and functional residual capacity (FRC) (PImax and PIFRC, respectively) and maximal expiratory mouth pressure was obtained at levels of total lung capacity (TLC) and FRC (PEmax and PEFRC, respectively). Patients performed maximal inspiratory and expiratory efforts against an obstructed mouthpiece at least three times at each lung volume and we adopted the mean value obtained from two reproducible values for analysis. The predicted values for age and sex correction were obtained from Black and Hyatt's formulae (1969), and percentages of PImax and PEmax for predicted values (%PImax and %PEmax, respectively) were calculated.

Slow vital capacity (VC) and forced expiratory volume in one second (FEV₁) were measured using a 13.5L Benedict-Roth type spirometer and the ratio of FEV₁ for VC (FEV₁/VC) was calculated. FRC was obtained with the He-gas dilution method (Meneely et al. 1960) using a water-sealed spirometer. TLC and RV were also obtained. %VC, %TLC, %FRC and %RV were calculated by Cotes' prediction (1979). Static pressure-volume curves were obtained with an esophageal balloon catheter system and body plethysmography according to standard techniques (Macklem et al. 1974). Maximum esophageal pressure (Pesmax) and static lung compliance (Cst) were also obtained (Macklem et al. 1974). Diffusing capacity for carbon monoxide (DLCO) was measured with the single breath
method (Forster 1957), and diffusing capacity per unit volume (DLCO/VA) was calculated. The single N₂ washout test was performed (Comroe and Fowler 1951), and closing volume (CV) and the ratio of closing capacity to TLC (CC%) were obtained. Two ml of arterial blood were sampled anaerobically from the brachial artery prior to measurements of all other respiratory function tests while the patient breathed room air, and the blood was quickly analyzed to obtain arterial oxygen tension (PaO₂), arterial carbon dioxide tension (PaCO₂) and pH with a pH blood gas analyzer (Model 213, Instrumentation Laboratories, Lexington, MA, USA). We defined hypoxemia as the state in which PaO₂ was below 80 torr, and hypercapnia as that when PaCO₂ was above 45 torr. All measurements of pulmonary function tests were performed in a sitting position and within approximately two hours.

**Statistical analysis**

All values are given as means ± S.D. Differences for each disease were evaluated by analysis of variance. Relationships between respiratory muscle functions and pulmonary functions were assessed by linear regression analysis and evaluated with the Student’s t-test. Values of p < 0.05 were considered to be statistically significant.

**RESULTS**

**Anthropometric data**

The anthropometric data are shown in Table 1. The age of the CPE patients was significantly higher and body weight of patients with IPF was significantly greater than the others. Height did not differ significantly among the three groups.

**Pulmonary functions**

Pulmonary functions are shown in Table 2. In NMD, %VC tended to be smaller and %RV and CC% tended to be larger than normal values (Cotes 1979). FEV₁%, %TLC, %FRC, P_{es\text{max}}, Cst and DLCO/VA were within normal ranges. Although mean values of PaO₂ and PaCO₂ were within normal ranges, PaO₂ varied from 55.1 to 93.7 torr and PaCO₂ from 30.4 to 78.9 torr. There were six patients with hypoxemia without hypercapnia. There was a significant inverse correlation between PaO₂ and PaCO₂ (PaCO₂ = -0.50 × PaO₂ + 79.5, r = -0.56,

<table>
<thead>
<tr>
<th>Table 1. Anthropometric data in neuromuscular diseases, chronic pulmonary emphysema and idiopathic pulmonary fibrosis</th>
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<td>N</td>
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<tr>
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</tr>
<tr>
<td>NMD</td>
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<tr>
<td>CPE</td>
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<td>IPF</td>
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NMD, neuromuscular diseases; CPE, chronic pulmonary emphysema; IPF, idiopathic pulmonary fibrosis.

Values shown are means ± S.D.

*P < 0.05 compared with other diseases.
Table 2. Pulmonary functions in neuromuscular disease, chronic pulmonary emphysema and idiopathic pulmonary fibrosis

<table>
<thead>
<tr>
<th></th>
<th>%VC (%)</th>
<th>FEV₁ (%)</th>
<th>%TLC (%)</th>
<th>%FRC (%)</th>
<th>%RV (%)</th>
<th>P_{esmax} (cmH₂O)</th>
<th>Cst (L/cmH₂O)</th>
<th>DLCO/VA (ml/min/mmHg/liter)</th>
<th>CC% (%)</th>
<th>PaO₂ (torr)</th>
<th>PaCO₂ (torr)</th>
</tr>
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<tbody>
<tr>
<td>NMD</td>
<td>81.9</td>
<td>79.5</td>
<td>98.6</td>
<td>97.3</td>
<td>128.7</td>
<td>19.3</td>
<td>0.25</td>
<td>7.2</td>
<td>49.5</td>
<td>76.8</td>
<td>41.4</td>
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<tr>
<td></td>
<td>±17.7</td>
<td>±8.6</td>
<td>±17.0</td>
<td>±10.4</td>
<td>±31.6</td>
<td>±3.6</td>
<td>±0.085</td>
<td>±1.4</td>
<td>±10.6</td>
<td>±12.7</td>
<td>±11.3</td>
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<tr>
<td>CPE</td>
<td>86.3</td>
<td>35.9*</td>
<td>121.4*</td>
<td>119.8*</td>
<td>172.6*</td>
<td>13.4*</td>
<td>0.39*</td>
<td>4.3*</td>
<td>68.3*</td>
<td>39.3</td>
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<tr>
<td></td>
<td>±18.3</td>
<td>±7.8</td>
<td>±12.1</td>
<td>±15.3</td>
<td>±35.3</td>
<td>±3.5</td>
<td>±0.14</td>
<td>±0.8</td>
<td>±8.6</td>
<td>±4.4</td>
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<tr>
<td>IPF</td>
<td>69.8</td>
<td>83.8</td>
<td>71.2*</td>
<td>73.5*</td>
<td>70.7*</td>
<td>52.3*</td>
<td>0.077*</td>
<td>5.1*</td>
<td>44.0</td>
<td>68.5*</td>
<td>36.7</td>
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<tr>
<td></td>
<td>±17.9</td>
<td>±5.4</td>
<td>±15.7</td>
<td>±11.7</td>
<td>±24.0</td>
<td>±18.6</td>
<td>±0.04</td>
<td>±1.6</td>
<td>±12.5</td>
<td>±6.2</td>
<td>±2.7</td>
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%VC, percent of vital capacity for predicted value; FEV₁,%, forced expiratory volume percent; %TLC, percent of total lung volume for predicted value; %FRC, percent of functional residual capacity for predicted value; %RV, percent of residual volume for predicted value; P_{esmax}, maximum esophageal pressure; Cst, static compliance; DLCO/VA, diffusing capacity for carbon monoxide per unit volume; CC%, a ratio of closing capacity to total lung volume percent; PaO₂, arterial oxygen tension; PaCO₂, arterial carbon dioxide tension; NMD, CPE and IPF are similar to Table 1.

Values shown are means ± s.d.

*p < 0.05 compared with neuromuscular disease.
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p < 0.05). In CPE, %TLC, %RV and Cst were increased, and FEV₁, %, Pesₘₐₓ, DLCO/VA and PaO₂ were decreased. In IPF, %VC and %TLC decreased and Pesₘₐₓ remarkably increased. Pulmonary function characteristics in the latter two groups were similar to those described by Fraser and Pare (1979).

Respiratory muscle functions are shown in Table 3. Mean PImax of patients with NMD was 44.6 ± 19.6 cmH₂O, which was significantly smaller than values in CPE and IPF (p < 0.01). %PImax in NMD was also significantly smaller than in the other two groups. %PImax in CPE (68.8 ± 20.5%) was significantly smaller than the value in IPF (p < 0.05). PIFRC, which is thought to be independent of respiratory recoil pressure showed the smallest values in NMD. PEMax of NMD was 59.6 ± 15.5 cmH₂O, which was significantly smaller than that in CPE and IPF. %PEmax of NMD was also smaller than that of the other two groups, which were approximately 60% of predicted values. PEFRC in NMD was smaller than that of the other two groups, in which PEFRC was within the normal range.

Relationship between respiratory muscle strength and pulmonary functions

The coefficient of correlation between %PImax or %PEmax and pulmonary function data obtained from linear regression analysis are shown in Table 4. In patients with NMD, there were significant positive correlations between %PImax and %TLC or %VC (r = 0.652 and r = 0.536, respectively), %PEmax and %TLC or %VC (r = 0.676 and r = 0.581, respectively) and %PImax and PaO₂ (r = 0.561), but no significant correlation between %PImax or %PEmax and PaCO₂. %RV was not correlated with %PEmax. In patients with IPF, %TLC, %VC, %FRC and %RV had a significant negative correlation with %PImax (r = −0.691, r =
Table 4. Correlation between respiratory muscle strength and pulmonary functions in neuromuscular diseases, chronic pulmonary emphysema and idiopathic pulmonary fibrosis

<table>
<thead>
<tr>
<th></th>
<th>%VC</th>
<th>FEV₁ %</th>
<th>%TLC</th>
<th>%FRC</th>
<th>%RV</th>
<th>Pes&lt;sub&gt;max&lt;/sub&gt;</th>
<th>Cst</th>
<th>DLCO/VA</th>
<th>CC%</th>
<th>PaO₂</th>
<th>PaCO₂</th>
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<td>NMD</td>
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<tr>
<td>%PImax</td>
<td>0.54</td>
<td>0.35</td>
<td>0.65</td>
<td>0.33</td>
<td>0.26</td>
<td>−0.20</td>
<td>−0.28</td>
<td>−0.11</td>
<td>0.02</td>
<td>0.56</td>
<td>0.10</td>
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<td></td>
<td>p &lt; 0.05</td>
<td>n.s.</td>
<td>p &lt; 0.02</td>
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<td>n.s.</td>
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<td>n.s.</td>
<td></td>
<td>p &lt; 0.05</td>
<td>n.s.</td>
</tr>
<tr>
<td>%PEmax</td>
<td>0.58</td>
<td>0.05</td>
<td>0.68</td>
<td>0.13</td>
<td>0.01</td>
<td>−0.20</td>
<td>−0.59</td>
<td>−0.24</td>
<td>0.18</td>
<td>0.40</td>
<td>0.16</td>
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<td></td>
<td>p &lt; 0.05</td>
<td>n.s.</td>
<td>p &lt; 0.02</td>
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<td>CPE</td>
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<tr>
<td>%PImax</td>
<td>0.33</td>
<td>0.49</td>
<td>0.03</td>
<td>−0.39</td>
<td>−0.46</td>
<td>−0.30</td>
<td>0.37</td>
<td>0.09</td>
<td>0.29</td>
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<td></td>
<td>n.s.</td>
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<tr>
<td>%PEmax</td>
<td>−0.48</td>
<td>−0.35</td>
<td>−0.34</td>
<td>−0.47</td>
<td>−0.26</td>
<td>−0.53</td>
<td>0.36</td>
<td>0.20</td>
<td>−0.52</td>
<td>−0.02</td>
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<td>IPF</td>
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<tr>
<td>%PImax</td>
<td>−0.63</td>
<td>0.52</td>
<td>−0.69</td>
<td>−0.67</td>
<td>−0.58</td>
<td>0.03</td>
<td>−0.37</td>
<td>0.23</td>
<td>0.01</td>
<td>−0.16</td>
<td>0.39</td>
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<tr>
<td></td>
<td>p &lt; 0.02</td>
<td>n.s.</td>
<td>p &lt; 0.01</td>
<td>p &lt; 0.01</td>
<td>p &lt; 0.05</td>
<td>n.s.</td>
<td>n.s.</td>
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<tr>
<td>%PEmax</td>
<td>−0.29</td>
<td>0.11</td>
<td>−0.21</td>
<td>−0.17</td>
<td>−0.01</td>
<td>0.016</td>
<td>0.10</td>
<td>0.14</td>
<td>−0.01</td>
<td>−0.03</td>
<td>0.50</td>
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<td></td>
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n.s., no significance. Abbreviations of pulmonary functions and three pulmonary diseases are defined similar to Tables 1, 2 and 3. Values show correlation coefficients between respiratory muscle strength and pulmonary functions.
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-0.629, r = -0.673 and r = -0.577, respectively), but no significant correlation with %PEmax. Furthermore, respiratory muscle strength had no correlation with both PaO2 or PaCO2. In patients with CPE, respiratory muscle functions did not have any significant correlation with pulmonary functions, including PaO2 and PaCO2.

Fig. 1 shows the relationship between %PImax and %VC in NMD and IPF. %VC showed a significant positive correlation with %PImax in NMD (%VC = 64.0 + 0.403 × %PImax, r = 0.536, p < 0.05). On the other hand, in IPF, %VC was inversely correlated with %PImax (%VC = 116.5 - 0.467 × %PImax, r = -0.629, p < 0.05). In both groups, a decrease in lung volume is thought to be remarkable according to the progression of the condition of the disease, while change in %PImax was inverse. Fig. 2 illustrates the significant relationship between %VC and %PEmax in NMD (%VC = 53.5 + 0.891 × %PEmax, r = 0.581, p < 0.05). The relationships between %TLC and %PImax or %PEmax were similar to those between %VC and %PImax or %PEmax. The lower both maximal inspiratory and expiratory mouth pressure were, the smaller the lung volume was.

Fig. 3 shows the relationships between %PImax and PaO2 in NMD (PaO2 =
Fig. 2. Relationship between %PEmax and %VC in patients with NMD. The continuous line (%VC = 53.5 + 0.891 × %PEmax) is the regression line. %VC is significantly correlated with %PEmax (r=0.581, p<0.05).

Fig. 3. Relationship between %PImax and PaO₂ in patients with NMD. The continuous line (PaO₂ = 63.4 + 0.303 × %PImax) is the regression line. PaO₂ is significantly correlated with %PImax (r=0.561, p<0.05).
63.4 + 0.303 \times \% P_{I\text{max}}, r = 0.561, p < 0.05). This means that PaO_2 decreases with decrease in inspiratory muscle strength.

**DISCUSSION**

There are methods for the evaluation of the respiratory muscle function; i.e., maximal mouth pressure (Black and Hyatt 1969), transdiaphragmatic pressure (Agostoni and Mead 1964), tension-velocity curve (Kikuchi et al. 1982), tension-time index (Bellemare and Grassino 1982), diaphragmatic electromyogram (Gross et al. 1979) and respiratory muscle oxygen consumption (Shindoh et al. 1987). In the present study, we used maximal inspiratory and expiratory mouth pressures to estimate inspiratory and expiratory muscle strength, because measurements of maximal mouth pressure could be performed easily, compared with the methods noted above. However, since actual lung volumes where measurements were performed were not determined, the influence of lung volume on maximal mouth pressure was not clear.

Since maximal mouth pressure consists of two parts; the pressure generated by respirator muscle and the recoil pressure of the respiratory system, the pure mouth pressure generated by the respiratory muscle should be obtained after correction for recoil pressure. However, in the present study, lung volume was not measured while the maximal mouth pressure was obtained. Therefore, both P_{I\text{max}} and P_{E\text{max}} were overestimated. The elastic recoil pressure at FRC level is thought to be zero. The relationships between P_{I\text{max}} or P_{E\text{max}} and pulmonary functions were similar to the relationship between P_{I\text{FRC}} or P_{E\text{FRC}} and pulmonary functions. In the present study, we present the former relationships, because we could use Black and Hyatt’s formulae (1969) for predicted values of P_{I\text{max}} and P_{E\text{max}}. Predicted values for Japanese are not yet available.

NMD is a syndrome characterized by striated muscle weakness of whole body, and the respiratory muscles, including the diaphragm, are also impaired. Inspiratory muscle strength in patients with NMD was approximately 45% of predicted value, and was smaller than in CPE and IPF patients. These data were comparable with those of previous reports (Black and Hyatt 1971; Kreizer et al. 1978; Braun et al. 1983). Lung disease itself associated with NMD might affect the respiratory muscle strength as shown in CPE and IPF in the present study. However, we found no abnormal shadows in chest roentgenograms of patients with NMD. Furthermore, since there were only two patients with FEV_1\% values between 60 and 70\%, implying a slight airflow obstruction, the influence of lung disease on the present results in NMD seems to be negligible.

Relationships between respiratory muscle functions and lung functions in NMD, \%VC and \%TLC were significantly correlated with both maximal inspiratory and expiratory mouth pressures. It is possible that these results are due to the length-tension relationship of the respiratory muscles. However, since significant obstructive and restrictive impairment of the lung were not suggested,
a more likely explanation is that expiratory and inspiratory muscle weakness causes a decrease in lung volume. These results support the findings of previous reports (Black et al. 1971; De Troyer et al. 1980). Data on the correlation between respiratory muscle weakness and FRC or RV are conflicting. If respiratory muscle weakness occurs, residual volume would increase, because the maximal expiratory effort would not be sufficient. However, we found no significant correlation between changes in FRC or RV and respiratory muscle strength, which is consistent with reports by Inkey et al. (1974) and De Troyer et al. (1980).

In patients with NMD, %Plmax was significantly correlated with PaO2. However, there was no significant correlation between respiratory muscle strength and PaCO2. Furthermore, there was no significant correlation between PaO2 and FEV1% or CC%. These results seem to differ from results obtained by Braun et al. (1983), who reported that respiratory muscle strength, especially below 50% predicted, were well correlated with PaCO2, but were not with PaO2. Although the reason for the discrepancy between Braun's and ours is not clear, a possible explanation may be the difference in the degree of respiratory dysfunction between the two studies.

The present results support the hypothesis that inspiratory muscle in sufficiency may cause hypoxemia (Roussos and Macklem 1977). There are several possibilities to explain the mechanisms. The first possible explanation is the increased unevenness of the ventilation-perfusion (VA/Q) ratio. Loh et al. (1979) showed that, in paralysis of the diaphragm, there was a marked decrease in the VA/Q ratio to the lower lung and suggested that the low VA/Q can contribute to posturally related hypoxemia. Campbell (1965) also stated that patients with motoneuron disease might exhibit hypoxemia in the presence of normal PaCO2. He speculated that in closed sites in the lung, nitrogen in the air, which is only slowly absorbed by the blood, is replaced by a gas which is rapidly absorbed, i.e., oxygen, and these parts of the lung ceased to be ventilated but might continue to be perfused, so that hypoxemia without hypercapnia might occur. Therefore, change in VA/Q ratio would be a most likely explanation in NMD patients with normocapnia. The second possible explanation is an alveolar hypoventilation due to respiratory muscle weakness. Since we observed a significant inverse correlation between PaO2 and PaCO2, this effect seemed to be significant, especially in patients with hypercapnia. The third possible explanation is microatelectasis due to inspiratory insufficiency. Gibson et al. (1977) showed that Cst is reduced according to respiratory muscle weakness because of lung microatelectasis. Since we observed no abnormality in Cst, microatelectasis in NMD patients studied may not have been significant.

In patients with IPF, %VC and %TLC had a significant inverse correlation with %Plmax. This is contrary to the data from NMD. A decrease in lung volume induces a decrease in the radius of curvature of the diaphragm. Therefore, an increase in transdiaphragmatic pressure may be explained by Laplace's
law. Another possible explanation is that inspiratory muscles may be subject to training effects from intrinsic elastic loading. Moreover, we found a decrease in the mean of maximal expiratory mouth pressure. This may be explained by a decrease in lung volume, resulting in a decrease in tension produced by expiratory muscle shortening. However, we found no correlation between expiratory muscle strength and lung volume. Furthermore, it is unlikely that respiratory muscle strength affects blood gas exchange in IPF.

We found a decrease in maximal inspiratory mouth pressure in CPE. This could be explained by hyperinflation of the lungs, resulting in a decrease in tension produced by inspiratory muscle shortening. However, we could find no correlation between maximal inspiratory mouth pressure and lung volume. Therefore, a decrease in inspiratory muscle strength may be affected by other factors; i.e., muscle fatigue, hypoxemia or malnutrition. However, blood gas was not correlated with respiratory muscle strength.

In summary, we found that inspiratory muscle strength had a significant correlation with %VC, %TLC and PaO₂ in NMD patients. These findings suggest that inspiratory muscle strength in NMD patients may be one important factor that causes hypoxemia and lung volume reduction. On the other hand, hypoxemia in CPE and IPF seemed to be determined independently by respiratory muscle strength.

References


