Lung Inflation Training Using a Positive End-expiratory Pressure Valve in Neuromuscular Disorders

Tsuyoshi Matsumura, Toshio Saito, Harutoshi Fujimura, Susumu Shinno and Saburo Sakoda

Abstract

Objective  Respiratory muscle weakness causes alveolar hypoventilation and reduced lung compliance in neuromuscular disorders. Lung inflation is important to prevent secondary pulmonary complications however respiratory and laryngeal dysfunction often hamper lung inflation. There is a need for a convenient and low-cost device that enables effective lung inflation. We tested a lung inflation training method using a positive end-expiratory pressure (PEEP) valve.

Methods  Vital capacity (VC), maximum insufflation capacity (MIC) and peak cough flow (PCF) as well as PEEP lung inflation capacity (PIC) were assessed in 93 neuromuscular patients. Consecutive PIC training was done for 4 months in six tracheostomized Duchenne muscular dystrophy (DMD) patients and PIC was assessed before and after training.

Results  PIC training was practicable in all participants and no serious adverse events were detected. PIC was significantly higher than VC or MIC in all disorders, although MIC was higher than VC in DMD only. Patients with dysphagia showed lower MIC and PCF compared with non dysphagic patients. PIC was as low as 345±77 mL in tracheostomized DMD patients; however consecutive training increased it up to 619±205 mL.

Conclusion  The PEEP valve enabled effective lung inflation regardless of laryngeal function. Consecutive training can improve lung condition even in advanced cases. Early introduction of PIC training could be effective at preventing respiratory complications in patients with neuromuscular disorders.

Key words: neuromuscular disorders, respiratory physical therapy, lung inflation training, positive end-expiratory pressure, resuscitation bag

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Materials and Methods

The institutional ethical review board of Toneyama National Hospital approved the protocol of this study, and all participants provided informed written consent.

Subjects were selected from patients with ALS, myotonic dystrophy (DM) or Duchenne muscular dystrophy (DMD), because respiratory dysfunction is inevitable in these disorders and bulbar dysfunction is common in ALS and DM. Those with obvious bullae, bronchial asthma, other active pulmonary illness, a history of pneumothorax, or severe congestive heart failure were excluded.

Single training effects were analyzed in 93 patients (ALS, 40; DM, 21; DMD, 32; Table 1) whose vital capacity (VC) and MIC (only non-tracheostomized patients) exceeded 50 mL. In this study, we measured VC, MIC (4), and PEEP lung insufflation capacity (PIC) in the supine position using a Wright/Haloscale respirometer. Peak cough flow (PCF) was also measured using a peak flow meter. Tracheostomized patients were excluded from MIC assessment, as air stacking is not possible in these patients. All measurements were made by respiratory therapists. We measured PIC by attaching the PEEP valve (20 cm H2O) to the resuscitation bag, tightly fitting the mask on the patient’s face (for tracheostomized patients, the resuscitation bag was connected to the tracheal cannula), delivering air until a leak started from the PEEP valve, releasing the PEEP valve, and inducing maximum expiration. We performed each procedure three times and recorded the maximum value. Because intraindividual variability in VC of these patients was less than 15%, we regarded MIC and/or PIC as effective when MIC or PIC exceeded 120% of VC, no change when MIC or PIC was between 80 and 120% of VC, and deteriorated when MIC or PIC was less than 80% of VC. To assess the effect of dysphagia (bulbar dysfunction) on lung inflation, patients were divided according to their diet; those without obvious dysphagia and eating an ordinary diet (ordinary group) and those with dysphagia and eating an arranged diet (thickening diet, blender diet, jelly etc.) or receiving tube feeding (dysphagic group).

Long-term effects were assessed in six tracheostomized DMD patients over 4 months; all six were inpatients with a mean age of 41.7±7.1 years and a mean duration of respiratory management of 13.7±2.9 years. They could hardly perform deep breathing and VC was all under 50 mL (Table 1). In terms of cardiac function, brain natriuretic peptide was 23.0±8.8 pg/mL, left ventricular diastolic diameter was 45.3±6.0 mm, and left ventricular ejection fraction was 35.3±8.0%. None showed obvious cardiac symptoms. Two sets of five lung inflations at 20 cm H2O of PEEP were done by nursing staff 5 days per week for 4 months, and the maximum PIC before and after the training were compared.

The number of febrile events due to respiratory infection in the 4 months before and during the study was also counted.

Statistical analysis

We used the paired t-test to compare VC, MIC, and PIC and to evaluate PIC before and after training; the Wilcoxon signed-rank test to compare the effect of MIC and PIC; analysis of covariance (ANCOVA) using VC as covariate to compare MIC, PIC, and PCF between the ordinary and

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**Table 1. Participants Profiles**

<table>
<thead>
<tr>
<th>n (M/F)</th>
<th>Age (y.o.) (mean ± SD)</th>
<th>DD (year) (mean ± SD)</th>
<th>VC (mL) (mean ± SD)</th>
<th>PCF (L/sec) (mean ± SD)</th>
<th>Alb (g/dL) (mean ± SD)</th>
<th>Diet (Ord/Arr/Tube)</th>
<th>Ventilation (none/NIV/TIV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALS 40 (19/21)</td>
<td>63.0 ± 12.4</td>
<td>3.5 ± 2.3</td>
<td>1200 ± 794</td>
<td>122 ± 92</td>
<td>3.05 ± 0.37</td>
<td>12/11/7</td>
<td>23/13/4</td>
</tr>
<tr>
<td>DM 21 (14/7)</td>
<td>51.7 ± 9.2</td>
<td>20.8 ± 10.3</td>
<td>1277 ± 566</td>
<td>177 ± 84</td>
<td>3.60 ± 0.37</td>
<td>16/5/0</td>
<td>10/11/0</td>
</tr>
<tr>
<td>DMD 32 (32/0)</td>
<td>18.0 ± 5.6</td>
<td>15.0 ± 4.6</td>
<td>1157 ± 636</td>
<td>172 ± 84</td>
<td>4.35 ± 0.49</td>
<td>30/2/0</td>
<td>16/16/0</td>
</tr>
<tr>
<td>TIV patients</td>
<td>Age (y.o.) (mean ± SD)</td>
<td>VC (mL) (mean ± SD)</td>
<td>PCF (L/sec) (mean ± SD)</td>
<td>Alb (g/dL) (mean ± SD)</td>
<td>Diet (none/NIV/TIV)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>ALS 4 (4/0)</td>
<td>61.8 ± 9.7</td>
<td>5.6 ± 3.1</td>
<td>425 ± 530</td>
<td>43 ± 75</td>
<td>3.53 ± 0.34</td>
<td>0/0/4</td>
<td>2.3 ± 2.8</td>
</tr>
<tr>
<td>DMD 6 (6/0)</td>
<td>41.7 ± 7.1</td>
<td>38.0 ± 6.5</td>
<td>&lt;50</td>
<td>unmeasurable</td>
<td>3.78 ± 0.56</td>
<td>2/2/3</td>
<td>13.7 ± 2.9</td>
</tr>
</tbody>
</table>

Abbreviations: M, male; F, female; y.o., years old; DD, disease duration; VC, vital capacity; PCF, peak cough flow; Alb, serum albumin; Ord, oral intake of ordinary diet; Arr, oral intake of arranged food (thickening diet, blender diet, jelly, etc.); Tube, tube feeding; NIV, non-invasive ventilation; TIV, tracheal intermittent positive pressure ventilation; MVD, duration receiving mechanical ventilation therapy.
Figure 1. Comparison of vital capacity (VC), maximum insufflation (MIC), and positive end-expiratory pressure (PEEP) insufflation capacity (PIC) in each disorder. Paired t-test was used to assess differences. In all disorders, PIC was higher than VC or MIC. In Duchenne muscular dystrophy (DMD), MIC was significantly higher than VC.

Results

Single training

The PIC training was practicable for all participants. PIC exceeded 400 mL in all subjects. VC, MIC, and PIC were 1,200±794, 1,226±868 and 1,644±727 mL, respectively for ALS patients; 1,277±566, 1,400±572 and 1,752±536 mL for DM patients; and 1,157±636, 1,441±762 and 1,689±815 mL for DMD patients. For DMD patients, MIC was significantly greater than VC, but no significant differences between MIC and VC were detected for ALS (p=0.693) and DM (p=0.131) patients. In all disorders, PIC was significantly greater than VC or MIC (Fig. 1).

Among non-tracheostomized patients, PIC training showed a higher rate of effectiveness than MIC training in all disorders, although significant differences were detected in only ALS (p=0.002) and DM (p=0.021) patients (Table 2). PIC was higher than 120% of MIC in 20 of 34 patients with ALS, 8 of 18 with DM and 13 of 30 with DMD. No subjects presented PIC under 80% of VC, although the MIC of nine patients was below 80% of VC. PIC was lower than 80% of MIC in one patient who complained strongly of discomfort and was reluctant to do PIC training.

VC was lower in the dysphagic group compared with the ordinary group, although differences were not significant. MIC, PIC, and PCF were compared with ANCOVA with VC as a covariate. As a result, MIC (p=0.001) and PCF (p=0.001) were significantly lower in the dysphagic group. Although PIC was also slightly lower in the dysphagic group, the difference was not significant (p=0.105) (Fig. 2).

No serious complications were detected during the study, although seven patients who did not receive mechanical ventilation and were unfamiliar with positive pressure complained of the difficulty in expiration that was caused by the mask and/or the PEEP valve.

Long-term training

The mean PIC of tracheostomized DMD patients was 345±77 mL, which was significantly lower than that of tracheostomized ALS patients (1,128±724 mL, p=0.027). In these patients, sustained PIC training for 4 months significantly increased PIC up to 619±205 mL (p=0.009). PIC was increased in all patients, although one patient with severe deformity (Kobb angle : 105°) showed minimum gain (Fig. 3A). No patient complained, and no obvious changes in vital signs (mean changes between before and after training was 0.3±5.1 mmHg in systolic blood pressure, -0.6±5.0 mmHg in diastolic blood pressure and 2.1±4.4/min in pulse rate) were observed during the training. The number of febrile events was decreased in three patients, although one patient presented with fever only after the start of training (Fig. 3B).
Lung inflation training is essential for NMD patients to prevent secondary lung diseases. Practicability, availability,
safety, portability and low cost are important factors in lung inflation. MIC is a common method, although many patients
cannot achieve adequate inflation because of laryngeal dys-
function or technical problems. In this study, the effect of
MIC was poor in patients with ALS and DM. Although MIE
is an effective tool, Japanese medical insurance allows its
use only for NMD patients receiving HMV. Thus it is not
suitable for patients who are not undergoing respiratory
management. It is noteworthy that PIC training meets most
of the requirements specified above. All equipment used in
this study is commercially available at a low cost, also it is
very compact and light, making it convenient and easily
transportable.

In this study, all subjects, including tracheostomized pa-

tients could perform PIC training. At first, we were con-

cerned that 20 cm H2O would be insufficient for lung infla-
tion because previous studies (5, 9, 12, 13) employed higher
pressure. Although there is another tool that allows for
higher pressure (14), this tool has not been approved as a
medical device in Japan. Despite our concern, PIC was sig-
ificantly higher than VC or MIC in all disorders. The rate
of effectiveness was also higher in PIC than MIC in ALS
and DM. No patients were assessed as deteriorated in PIC
compared to VC. These facts confirm that PIC is an effec-
tive technique for lung inflation and independent from la-
ryngeal function or techniques.

No serious adverse events were detected during the single
or long-term study. Vital signs were not influenced even in
advanced cases. We believe the safety of PIC training is
high, although it is important to exercise caution in patients
with contraindications such as active pneumothorax, or se-
vere congestive heart failure.

PIC training was less effective when patients resisted it.
PIC was less than 80% of MIC in a patient who was reluc-

Table 2. Cross-tabulation Table Showing Effectiveness of MIC and PIC

<table>
<thead>
<tr>
<th></th>
<th>Effective</th>
<th>No change</th>
<th>Deteriorated</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>PIC</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>ALS (n=34)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Effective</td>
<td>9</td>
<td>10</td>
<td>3</td>
</tr>
<tr>
<td>No change</td>
<td>2</td>
<td>9</td>
<td>1</td>
</tr>
<tr>
<td>Deteriorated</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>DM (n=18)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Effective</td>
<td>8</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>No change</td>
<td>1</td>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>Deteriorated</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td><strong>DMD (n=30)</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Effective</td>
<td>16</td>
<td>4</td>
<td>1</td>
</tr>
<tr>
<td>No change</td>
<td>1</td>
<td>6</td>
<td>2</td>
</tr>
<tr>
<td>Deteriorated</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

MIC, maximum insufflations capacity; PIC, positive end-expiratory pressure inflation capacity; ALS, amyotrophic lateral sclerosis; DM, myotonic dystrophy; DMD, Duchenne muscular dystrophy.

Each subgroup was determined as follows: effective, value exceeded 120% of VC; no change, the value was within 80-120% of VC; deteriorated, value was below 80% of VC.

**Discussion**

Lung inflation training is essential for NMD patients to prevent secondary lung diseases. Practicability, availability,
important to train. Some patients who were not on mechanical ventilation complained about the discomfort associated with PIC training. In such cases, an adequate explanation and relaxation are important to maximize the effects.

PIC was low in six tracheostomized DMD patients. Although not significant, dysphagic patients showed lower PIC than non-dysphagic patients. These findings suggest that lung compliance may have been damaged in patients with advanced disease. Higher pressure might enable more inflation, although it is critical to avoid barotrauma (10, 11). To date, both positive (5, 9) and negative (12, 13) findings have been reported for the effect of positive pressure training, and it is suggested that a brief period of lung inflation may not be enough to reverse long-standing alterations in the lung (12, 15). Thus we assessed the long-term effect of the PIC training for tracheostomized DMD patients. Similar to a recent report (8), PIC was improved in all patients with sustained training. In addition, febrile events were also reduced. It seems that consecutive PEEP lung inflation is effective at improving lung conditions even in advanced cases.

**Conclusion**

Lung inflation training with a PEEP valve is effective, convenient, and safe in NMD patients. Although consecutive training was effective even in advanced cases, we recommend early preventive introduction of this training. We hope that wider use of this lung inflation method will improve the health and quality of life of NMD patients.

**The authors state that they have no Conflict of Interest (COI).**

**Acknowledgement**

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**References**