**Objective:** To investigate the effect of deep lung insufflations on maximum insufflation capacities (MICs) and peak cough flows (PCFs) for patients with neuromuscular disease.

**Method:** Forty-three patients with neuromuscular disease were trained in stacking delivered volumes of air to deep lung insufflation and were prescribed a program of air stacking once their vital capacities (VCs) were noted to be < 2,000 mL. VC, MIC, and unassisted and assisted PCF were monitored. The initial data were compared with the highest MICs subsequently achieved. For those patients whose MICs only decreased, we compared the initial data with the most recent data.

**Results:** The MICs increased from (mean ± SD) 1,402 ± 530 mL to 1,711 ± 599 mL (p < 0.001) for 30 patients and only decreased for 13 patients. Patients for whom the MICs increased also had a significant increase in assisted PCF from 3.7 ± 1.4 to 4.3 ± 1.6 L/s (p < 0.05) despite having somewhat decreasing VCs and unassisted PCFs.

**Conclusion:** With training, the capacity to stack air to deep insufflations can improve despite progressive neuromuscular disease. This can result in increased cough effectiveness.

**Key words:** cough; maximum insufflation capacity; neuromuscular disease; peak cough flow; pulmonary compliance; range-of-motion therapy

**Abbreviations:** ALS = amyotrophic lateral sclerosis; DMD = Duchenne muscular dystrophy; MIC = maximum insufflation capacity; PCF = peak cough flows; ROM = range of motion; SMA = spinal muscular atrophy; VC = vital capacity

Neuromuscular diseases are characterized by progressive weakening of skeletal, respiratory, bulbar, and, in the case of myopathies, cardiac muscles. Diagnoses are made on the basis of electrodiagnosis, muscle or nerve biopsies, elevations of serum muscle enzymes, and, in the case of Duchenne muscular dystrophy (DMD), spinal muscular atrophy (SMA), and a few rare conditions, by cellular DNA analysis. The inability to effectively cough out airway secretions is the main cause of respiratory failure and death for these patients.

Normal breathing consists of varying tidal volumes with intermittent deep breaths or sighs. Periodic hyperinflation is required to prevent closure of lung units. Patients with neuromusculoskeletal disease can have severe inspiratory and expiratory muscle weakness that diminishes tidal volumes, sighs, and cough flows, resulting in little expansion of the lungs and chest walls. This leads to stiffening of the rib cage and a reduction in chest wall compliance. Reduction in lung compliance and in lung and chest wall range of motion (ROM) also may be related to alterations in the elastic properties of lung tissues caused by the diminished lung volumes.

During a normal cough, about (mean ± SD) 2.3 ± 0.5 L of air is expelled at flows of 6 to 20 L/s after glottic closure of about 0.2 s. High thoracoabdominal pressures are needed to generate effective cough flows on glottic opening. Thus, an effective cough requires deep lung volumes and breath holding. A deep breath dilates the airways, increases the force of expiratory muscle contraction, and increases lung recoil pressure. Breath holding facilitates the distribution of air to the lung periphery and causes intrathoracic pressure to increase. Thus, it is not surprising that assisted peak cough flows (PCFs) can be significantly increased from maximal insufflations.

The maximum insufflation capacity (MIC) is a function of oropharyngeal and laryngeal muscle function and, to some degree, of pulmonary compliance. We hypothesized that patients with neuromusculoskeletal disease who have diminished vital capacities (VCs) but adequate bulbar muscle function...
tion could benefit from training in and prescription of a daily program of air stacking to maximal lung insufflation, increasing MICs and assisted PCFs. The essential purpose of this study was to investigate the effect of lung ROM therapy on MICs and assisted PCFs.

MATERIALS AND METHODS

All patients with neuromusculoskeletal disease diagnosed by standard criteria and monitored in a Jerry Lewis Muscular Dystrophy Association clinic were candidates for this study, provided that they were old enough to cooperate. The exclusion criteria for taking part in the deep insufflation program were the following: VC > 2,000 mL; the presence of concomitant intrinsic lung disease to the extent of necessitating oxygen therapy; bulbar muscle dysfunction so severe that glottic closure could not be attained and the MIC, therefore, could not exceed the VC; the presence of an indwelling tracheostomy tube; the inability to cooperate because of mental retardation; and failure to return to the clinic for reevaluation. Chronic lung disease was defined by a baseline oxyhemoglobin saturation that is chronically less than 95% despite eucapnia and the absence of airway mucus accumulation, or an FEV1/FVC ratio < 2 SDs of normal.

The pulmonary function testing was performed by a respiratory therapist who had no idea that the data would be used for a study. The data analysis was retrospective, and the study was conceived after all of the data had been collected.

The patients were trained in air stacking in the clinic and were prescribed 10 to 15 maximal lung insufflations by air stacking three times each day from the point at which their VCs were noted to be < 2,000 mL. The VC, MIC, and unassisted and assisted PCFs were evaluated, and patients within 100 miles of the clinic were asked to return for reevaluations every 6 months. The MIC by air stacking was achieved by the patient taking a deep breath, holding it, and then air stacking consecutively delivered volumes of air to the maximum volume that could be held with a closed glottis. The air was delivered from a manual resuscitator or portable volume ventilator via a mouthpiece or nasal interface. The patient then exhaled the maximally held volume of air (MIC) into a Wright spirometer (model Mark 14; Ferraris Development and Engineering Co, Ltd; London, UK), and both the VC and the MIC were measured. The maximum values that were observed in four or five attempts were recorded.

Unassisted PCFs were measured by having the patient cough as forcibly as possible through a peak flowmeter (Assess; Health Scan Products Inc; Cedar Grove, NJ). To measure assisted PCFs, the patient was insufflated to a deep insufflation and then asked to cough forcefully through the peak flowmeter as an abdominal thrust was timed to glottic opening. The maximum observed flows in four or five attempts were recorded. Forced expiratory flows other than cough flows were also measured (Microspiro HI-501; Chest Corp; Tokyo, Japan).

The data at the initiation of the program and the values at the reevaluation at which the MICs were the highest were compared by paired t test. Values are given as mean ± SD. For those patients whose MICs only decreased after the initial evaluation, we compared the data from the initial evaluation with those of the most recent reevaluation. All patients were in either the increasing MIC group or the decreasing MIC group. The VC, MICs, and assisted PCFs of these groups also were compared with those of the MIC-equals-VC group. Longitudinal data and intergroup comparisons were made by t test. All p values < 0.05 were considered to be statistically significant.

RESULTS

One hundred eight patients with neuromuscular weakness underwent initial evaluations. Forty-three patients returned for reevaluation and satisfied the inclusion criteria. The 108 patients had the following diagnoses: DMD, 32 patients; amyotrophic lateral sclerosis (ALS), 30 patients; SMA, 15 patients; post-polioymyelitis syndrome, 12 patients; non-DMD myopathies, 14 patients; and miscellaneous, 5 patients. Two of the 108 patients had tracheostomy tubes. One patient did not return to clinic. Thirteen patients were unable to cooperate or were too young for the study. Thirty-one patients had VCs > 2,000 mL. Eighteen of the 108 patients were eliminated from the program because their MICs could not exceed their VCs. These 18 patients had the following diagnoses: ALS, 15 patients; SMA, 1 patient; multiple sclerosis, 1 patient; and non-Duchenne-type muscular dystrophy, 1 patient.

Of the 43 study patients, the 30 in the increasing MIC group had the following diagnoses: DMD, 9 patients (age, 22.5 ± 5.3 years); ALS, 6 patients (age, 53.9 ± 11.6 years); SMA, 3 patients (age, 11.3 ± 6.1 years); post-polioymyelitis syndrome, 6 patients (age, 61.1 ± 10.2); and miscellaneous, 6 patients (including non-Duchenne-type muscular dystrophy, 4 patients; congenital myopathy, 1 patient; and kyphoscoliosis, 1 patient; age, 33.1 ± 20.7 years). The 13 patients in the decreasing MIC group had the following diagnoses: DMD, 5 patients (age, 21.6 ± 2.3 years); ALS, 3 patients (age, 49.7 ± 16.5 years); SMA, 3 patients (age, 11.2 ± 4.0 years); and multiple sclerosis, 1 patient, and non-Duchenne-type muscular dystrophy, 1 patient, 48 and 28 years old, respectively. The relative functional abilities of the three groups of patients can be seen in Table 1. The increasing MIC group had significantly greater muscle strength than the decreasing MIC group (p < 0.05) at the data end point but not at the initial evaluations.

All 43 patients reported performing the air stacking at least twice daily; however, there was no way to confirm this. The changes in VCs, MICs, and PCFs with the insufflation program are given in Table 2 and Figure 1. None of the initial values shown in Table 2 were significantly different between the increasing MIC group and the decreasing MIC group. Of the 30 ALS patients who were evaluated and the 18 neuromuscular disease patients whose MICs did not exceed their VCs because of severe bulbar muscle involvement, 15 of the patients had ALS. The VC-equals-MIC patients, despite having significantly greater VCs and general muscle strength, had significantly lower PCFs (p < 0.01) than the MC-greater-than-VC groups.
The lung and chest wall is usually ignored, and when combined with ROM therapy for the extremities, the ROM of the lung and chest wall can be maintained.16,17 Lung and chest wall ROM can be maintained limb mobility (joint mobilization) for patients with muscular weakness. The loss of limb function results from weakness as well as from musculotendinous tightness or contractures.16 Contracture prevention or correction can help to maintain function.16,17 Lung and chest wall ROM can be provided by giving deep insufflations. By contrast with ROM therapy for the extremities, the ROM of the lung and chest wall is usually ignored, and when this results in submaximal cough flows, it can be life-threatening. In this study, pressure-limited ventilators were not used because the flow pressures of the available models cannot fully insufflate the lungs.

It could not be confirmed how well the patients complied with the prescribed insufflation therapy. Assuming that none of them performed any regular insufflation therapy and practiced this technique only during clinic visits with us, the importance of the results of this study are not diminished. Whether the increase in air-stacked volumes was due to a training effect of practice at home or occurred only in our clinic, it is the extent of the increase in MIC that correlates with increases in assisted cough flows and, therefore, the ability to cough effectively. The higher the cough flow, the less likely are respiratory complications of neuromuscular disease.

Hypercapnia results from decreased tidal volumes due to a combination of inspiratory muscle weakness and an increase in the dynamic elastance of lung tissues.18 The latter apparently results from an inability to take the deep breaths needed to maintain pulmonary compliance. Long-term failure to take deep breaths results in chronic microatelectasis and loss of lung and chest wall elasticity with decreased static pulmonary compliance.19–21 As a consequence, the MIC becomes smaller along with the VC. We have observed patients who have been chronically underinflated for years who with only a 100-mL increase in ventilator-delivered volumes had painful stretching of intercostal musculature. In this study, we demonstrated the ability to significantly increase lung and chest wall ROM (MICs) and PCFs for the majority of patients with neuromusculoskeletal conditions despite their having severe and progressive generalized, inspiratory, and bulbar muscle weakness. In some cases, increases in MIC and assisted PCF were noted despite decreasing VC and unassisted PCF. Some of our patients with little or no measurable VC have maintained MICs of ≥ 2 L for decades with ongoing lung insufflation therapy. It is unclear whether increases in MIC reflect improvement in pulmonary compliance or are the effect of practice and better control of bulbar musculature.

### Table 1—Group Comparisons of Strength and Function

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Increasing MIC (n = 30)</th>
<th>Decreasing MIC (n = 13)</th>
<th>MIC = VC (n = 18)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, yr</td>
<td>36.1 ± 21.0</td>
<td>28.2 ± 16.7</td>
<td>54.3 ± 16.8</td>
</tr>
<tr>
<td>Dysphagia</td>
<td>7</td>
<td>3</td>
<td>11</td>
</tr>
<tr>
<td>Follow-up</td>
<td>9</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Dysarthria</td>
<td>2</td>
<td>3</td>
<td>11</td>
</tr>
<tr>
<td>Follow-up</td>
<td>2</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Gastrostomy tube</td>
<td>2</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Follow-up</td>
<td>3</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>Wheelchair at initial point</td>
<td>21</td>
<td>12</td>
<td>10</td>
</tr>
<tr>
<td>and follow-up</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PT ventilator use†</td>
<td>12</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Initial point</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>14</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>FT ventilator use‡</td>
<td>9</td>
<td>2</td>
<td>0</td>
</tr>
<tr>
<td>Initial point</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Follow-up</td>
<td>10</td>
<td>3</td>
<td></td>
</tr>
<tr>
<td>MMT§ mean score</td>
<td>1.7 ± 1.4</td>
<td>1.5 ± 1.4</td>
<td>2.3 ± 1.5</td>
</tr>
</tbody>
</table>

*Values are given as mean ± SD or No. PT = part-time; FT = full-time; MMT = manual muscular testing.
†Part-time defined as 8 to 16 h of ventilator use.
‡Full-time defined as 20 to 24 h of ventilator use.
§Manual muscle testing is the averaging of major muscle groups (upper limb shoulder abductors, elbow flexors, wrist extensors and lower limb hip flexors, quadriceps, and ankle dorsiflexors).

### Discussion

Extremity ROM therapy is a cornerstone of physical medicine and rehabilitation interventions to maintain limb mobility (joint mobilization) for patients with muscular weakness. The loss of limb function results from weakness as well as from musculotendinous tightness or contractures.16 Contracture prevention or correction can help to maintain function.16,17 Lung and chest wall ROM can be provided by giving deep insufflations. By contrast with ROM therapy for the extremities, the ROM of the lung and chest wall is usually ignored, and when

### Table 2—Outcome of Maximum Insufflation Program

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Increasing MIC Group (n = 30)</th>
<th>Decreasing MIC Group (n = 13)</th>
<th>MIC = VC Group (n = 18)</th>
</tr>
</thead>
<tbody>
<tr>
<td>MIC, mL</td>
<td>1,403 ± 530</td>
<td>1,711 ± 599</td>
<td>1,276 ± 474</td>
</tr>
<tr>
<td>VC, mL</td>
<td>904 ± 474</td>
<td>895 ± 432</td>
<td>1,236 ± 327</td>
</tr>
<tr>
<td>APCF, L/s</td>
<td>3.7 ± 1.4</td>
<td>4.3 ± 1.6</td>
<td>1.5 ± 1.4</td>
</tr>
<tr>
<td>UPCF, L/s</td>
<td>2.5 ± 1.6</td>
<td>2.4 ± 1.5</td>
<td>1.5 ± 1.5</td>
</tr>
</tbody>
</table>

*Values given as mean ± SD. APCF = assisted PCF; UPCF = unassisted PCF. Initial = initiation of insufflation program; Highest = maximum MIC value noted during follow-up; Last = last reevaluation.
Cough flows can decrease from both inspiratory and expiratory muscle weakness. Incomplete or weak glottic closure can exacerbate cough dysfunction and further decrease PCFs.22 Cough flows < 160 L/min are ineffective.23 Assisted coughing, including air stacking, usually can increase PCFs to a point over the 160 L/min threshold.10 In 1966, Kirby and colleagues12 demonstrated a mean increase in PCFs from 3.6 to 6.5 L/s for traumatic tetraplegic patients with cough assistance. More recently, assisted coughing has been reported to be instrumental in averting episodes of respiratory failure, hospitalization, and the need to resort to tracheostomy for patients with DMD.24 Studies of nocturnal nasal ventilation for patients with neuromuscular disease that did not include air stacking and assisted coughing during intercurrent chest colds failed to significantly delay a resort to tracheostomy or death.25–28

The group with MICs equal to VCs had the most severely dysfunctional bulbar musculature despite having significantly greater skeletal muscle strength and VCs than the groups with MICs greater than VCs. This situation is often seen in ALS patients and is the reason that so many of these patients eventually need to undergo a tracheostomy to prolong their survival. The patients with decreasing MICs had greater bulbar muscle function than the group with MICs equal to VCs, but less than the increasing MIC group. This can be seen because their MICs exceed their VCs but fail to increase, their assisted PCFs are significantly less than those of the increasing MIC group, and they had a higher percentage of patients who developed severe dysphagia and dysarthria and required gastrostomy tubes. Their skeletal muscle strength also was not significant than and deteriorated faster than that of the increasing MIC group, so that they appeared to have a more rapid disease course. Since none of the study patients had COPD or intrinsic lung disease and since the decrease in MIC in the decreasing MIC group was not related to VC, it probably was due to bulbar muscle dysfunction and possibly, to some degree, to decreases in pulmonary compliance. On the other hand, we have found that patients with normal bulbar muscle function, such as those with spinal cord injury, can improve and maintain MICs and assisted PCFs with practice.10 In 1980, Huldgren et al29 first demonstrated that maximal insufflation therapy could increase MIC after spinal cord injury.

In conclusion, the MICs of the majority of patients (30 of 43 [70%]) with neuromuscular disease can increase, resulting in significantly increased assisted PCF. The most important exception is that of bulbar ALS patients.

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