Use of the Mechanical In-Exsufflator in Pediatric Patients With Neuromuscular Disease and Impaired Cough*

Laura J. Miske, MSN; Eileen M. Hickey, CRNP; Susan M. Kolb, CRNP; Daniel J. Weiner, MD, FCCP; and Howard B. Panitch, MD, FCCP

Background: Impaired cough secondary to weakness from neuromuscular disease (NMD) can cause serious respiratory complications, including atelectasis, pneumonia, small airway obstruction, and acidosis. The mechanical in-exsufflator (MI-E) delivers a positive-pressure insufflation followed by an expulsive exsufflation, thereby simulating a normal cough. Use of the MI-E in adults with impaired cough results in improved cough flows and enhanced airway clearance. However, only limited reports of MI-E use in children exist.

Objective: To determine the safety, tolerance, and effectiveness of the MI-E in a pediatric population.

Method: Retrospective medical record review.

Participants: Sixty-two patients (34 male patients) observed in a pediatric pulmonary program with NMD and impaired cough in whom MI-E therapy was initiated. Median age at initiation of MI-E use was 11.3 years (range, 3 months to 28.6 years). Diagnoses included the following: Duchenne muscular dystrophy (17 patients); spinal muscular atrophy, types I and II (21 patients); myopathy (12 patients); other nonspecific NMD (12 patients). Mechanical ventilation via tracheostomy was used in 29 patients, and 25 patients used noninvasive ventilation.

Results: The median duration of use was 13.4 months (range, 0.5 to 45.5 months). One infant died before using MI-E at home. Five patients chose not to continue MI-E therapy. Complications were reported in two patients, but ultimately they used the MI-E device. Chronic atelectasis resolved in four patients after beginning MI-E therapy, and five patients experienced a reduction in the frequency of pneumonias.

Conclusion: In 90% of our study population, the use of an MI-E was safe, well-tolerated, and effective in preventing pulmonary complications.

Key words: airway clearance; cough; cough peak flow; Duchenne muscular dystrophy; mechanical in-exsufflator; neuromuscular disease; pediatrics; spinal muscular atrophy

Abbreviations: ANOVA = analysis of variance; CPF = cough peak flow; DMD = Duchenne muscular dystrophy; MI-E = mechanical in-exsufflator; MYO = congenital myopathy; NMD = neuromuscular disease; NS-NMD = nonspecific neuromuscular disease; Pemax = maximal expiratory pressure; Pinmax = maximal inspiratory pressure; SMA = spinal muscular atrophy

With aggressive ventilatory support, the life expectancy of children with neuromuscular disease (NMD) has been extended beyond initial expectations.1 The application of positive or negative pressure ventilation can reverse respiratory failure resulting from progressive muscle weakness and chest wall distortion. Children with moderate or severe respiratory muscle weakness, however, are still at risk for recurrent episodes of pneumonia or atelectasis because of an inability to clear the airways of secretions. Ineffective airway clearance can be exacerbated by acute respiratory illnesses, when secretion production increases and respiratory muscle function can acutely deteriorate.2

An effective cough requires an inspiration to 60 to 80% of total lung capacity followed by glottic closure and a pause at the raised lung volume to accentuate the distribution of air throughout airways and beyond secretions.3 Intrathoracic pressure increases as...
expiratory muscles contract, and when the glottis is opened, air is expelled, and secretions are propelled toward the central airways and mouth. Each of these facets of a normal cough can be impaired in patients with neuromuscular weakness. When present, inspiratory muscle weakness limits the depth of the precough inspiration, bulbar weakness or presence of a tracheostomy impairs glottic closure, and expiratory muscle weakness or chest wall distortion from scoliosis reduces intrathoracic expiratory pressures and flows. Respiratory complications of ineffective airway clearance include pneumonia, atelectasis, and altered gas exchange, resulting in supplemental oxygen dependency and respiratory acidosis.4

Measures to improve airway clearance can minimize or prevent life-threatening conditions that typically require hospital admission for a patient with NMD. Both manual and mechanically assisted cough, with or without insufflation, have been used routinely in adult patients with NMD to avoid pulmonary morbidity and mortality.5-10 There are, however, limitations to manually assisted cough. It may not be effective in patients with chest wall distortion from scoliosis because of the difficulty in establishing optimal hand placement on the chest to enhance expiratory flows.5 Also, it must be performed cautiously in young infants and children with NMD, whose chest walls are abnormally compliant.11 Manual techniques are also time-intensive and require special training to optimize caregiver-patient coordination.

In the early 1950s, mechanical devices that provided a large inspiration followed by an exsufflation with negative pressure were used to treat patients with poliomyelitis and impaired cough.9,12,13 Similar mechanical insufflator-exsufflatators (the In-Exsufflator or CoughAssist; J.H. Emerson Co; Cambridge, MA) have subsequently been used successfully in adult patients with other types of neuromuscular weakness7,9,14 to assist with airway clearance as an interventional and preventive measure. There are, however, few reports13,15 of the use of mechanical cough assist devices in pediatric patients. The purpose of this study was to describe our experience with a cough assist device (the Mechanical In-Exsufflator [MI-E]) in a population of pediatric patients with NMD who were observed in the pulmonary program of a tertiary care center.

**Materials and Methods**

We began using the MI-E routinely for patients with impaired cough in our institution in November 1998. After approval by the institutional review board, we performed a retrospective medical record review of patients with NMD who had been followed in the Pulmonary Division of the Children’s Hospital of Philadelphia from that time until April 30, 2001. All patients to whom an MI-E device was prescribed for use at home were included in the study. Demographic information, including diagnosis, gender, age at initiation of use of the MI-E, use of antireflux medications and/or presence of fundoplication, presence of tracheostomy, and need for mechanical ventilatory support were recorded. In addition, to characterize the pulmonary impairment of this population, pulmonary function data from patients > 6 years of age, including spirometry and measurements of respiratory muscle strength from the most recent routine office visit before the initiation of the MI-E were noted as well. Data then were collected to determine the safety, tolerance, and efficacy of the MI-E in this patient population. Safety was assessed by noting the occurrence of pulmonary, cardiac, or GI complications (eg, pneumothorax, pulmonary hemorrhage, cardiac dysrhythmias, nausea, or vomiting) associated with use of the device. Patients were considered to tolerate the MI-E if they reported their use of the device at the prescribed frequency. They were judged not able to tolerate the device if they or their caregiver expressed the desire to discontinue the use of the MI-E for any reason. Efficacy was assessed by patient report related to secretion clearance and resolution of illness, or improvement in chest radiographs.

**Patient Use of the MI-E**

Patients were identified for use of the MI-E if their maximal expiratory pressure (P_{max}) was < 60 cm H_{2}O if there was a history of lower respiratory tract infection or atelectasis, or if they were weak enough to require long-term mechanical ventilatory support. The initial treatment with the MI-E was performed by respiratory therapists. Patients were evaluated in the outpatient office setting (32 patients) or in the acute hospital setting (30 patients) if they had been admitted for an acute respiratory illness. Clinical examination determined the pressure settings, as follows. Positive inspiratory pressures were established by visual inspection of chest wall excursion and auscultation for bilateral adequacy of air entry. Negative pressures were set at −15 cm H_{2}O and increased incrementally according to the patient’s comfort and production of secretions up to −40 cm H_{2}O. The positive-pressure breath was delivered to the patient over 2 to 3 s, while the negative exsufflation was delivered in 1 to 2 s. The breaths were coordinated with the patient’s own breath rate and rhythm. Three to five breaths were delivered followed by a period of rest before continuing, for a total of three to five cycles. Interfaces included a mouthpiece (6 patients) [Fig 1], facemask (25 patients), or direct connection to the tracheostomy tube (28 patients). Airway suctioning was performed at the end of each cycle and as needed during each rest period. Oxygen saturations were monitored during the initial trial and as necessary during subsequent treatments. Supplemental oxygen was administered through the device as necessary to maintain oxyhemoglobin saturations of > 94%.

**Results**

We identified 62 patients during the study period who used the MI-E. All patients had NMD and fell into the following categories: Duchenne muscular dystrophy (DMD); congenital myopathies (MYOs); nonspecific NMD (NS-NMD); and spinal muscular atrophy (SMA), type I and type II. Their characteristics are shown in Table 1.

Diagnoses included a wide variety of neuromuscular disorders, the age at onset of respiratory im-

---

**Table 1**

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>Number of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duchenne muscular dystrophy (DMD)</td>
<td>25</td>
</tr>
<tr>
<td>Congenital myopathies (MYOs)</td>
<td>10</td>
</tr>
<tr>
<td>Nonspecific NMD (NS-NMD)</td>
<td>15</td>
</tr>
<tr>
<td>Spinal muscular atrophy (SMA), type I and type II</td>
<td>12</td>
</tr>
</tbody>
</table>

---
pairment of which also typically varied from infancy (e.g., SMA type I) to adolescence (e.g., DMD). Thus, the range of ages at introduction of MI-E use for all subjects was wide (range, 0.25 to 28.6 years; median, 12.6 years). As expected, age at initiation of therapy varied by diagnosis (Table 1). For the group, 8 patients did not use any form of mechanical ventilatory support, 25 depended on noninvasive positive-pressure ventilation, and 29 used invasive ventilation via tracheostomy.

A total of 32 patients underwent pulmonary function testing during a routine office visit prior to the initiation of MI-E therapy (DMD, 10 patients; MYO, 6 patients; NS-NMD, 6 patients; and SMA type II, 10 patients) [Table 2]. For all patients, pulmonary functions were consistent with moderate-to-severe restrictive lung disease. In addition, respiratory muscle strength was diminished. Patients with DMD had significantly lower FVC and FEV₁ values compared with the other groups, as determined by one-way analysis of variance (ANOVA) [p < 0.01]. The maximal inspiratory pressure (Pmax) was different among the groups as well (p < 0.05), while all other measurements were similar among the four diagnostic groups.

Age and underlying cause of neuromuscular weakness were independent of pressures used with the MI-E device. There was no correlation between age and inspiratory pressure used (p = 0.07; r² = 0.23), or between age and expiratory pressure used (p = 0.18; r² = 0.18). Nor was there any relationship between diagnosis and inspiratory pressure used.

### Table 1—Patient Characteristics

<table>
<thead>
<tr>
<th>Diagnosis</th>
<th>DMD (n = 16)</th>
<th>MYO (n = 12)</th>
<th>NS-NMD (n = 12)</th>
<th>SMA Type I (n = 8)</th>
<th>SMA Type II (n = 14)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Median age at start, yr</td>
<td>21.0 (16.7–27.7)</td>
<td>10.7 (0.2–22.9)</td>
<td>9.7 (0.3–18.5)</td>
<td>2.4 (0.4–15.9)</td>
<td>12.6 (1.5–28.6)</td>
</tr>
<tr>
<td>Male patients</td>
<td>16</td>
<td>6</td>
<td>7</td>
<td>0</td>
<td>5</td>
</tr>
<tr>
<td>Noninvasive</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ventilation</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Tracheostomy with</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ventilation</td>
<td>11</td>
<td>7</td>
<td>5</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>No ventilatory support</td>
<td>0</td>
<td>1</td>
<td>3</td>
<td>2</td>
<td>2</td>
</tr>
</tbody>
</table>

*Values in parentheses are ranges.
(p = 0.981, by ANOVA on ranks) or expiratory pressure used (p = 0.983, by ANOVA on ranks). The median inspiratory and expiratory pressures for all groups were 30 and 30 cm H2O, respectively, with a range of pressures from 15 to 40 cm H2O (for insufflation) and 20 to 50 cm H2O (for exsufflation).

The frequency of reported use ranged from once a day to every 4 h on a routine basis, with most patients using the device twice daily. Patients who routinely produced secretions preferred using the device more frequently than twice daily. During periods of wellness, patients and caregivers, in consultation with the physician, adjusted the treatment schedule to meet their needs. Performing the maneuvers in the morning on awakening and again before bedtime seemed to work well for the majority of families in maintaining technique and effective airway clearance. Patients and caregivers who tried to use the device only during periods of a respiratory exacerbation found that they were not as comfortable with the device and were not as effective with airway clearance as they would have liked. Therefore, their treatment orders were changed to daily use of the MI-E device.

For 90% of the group, MI-E use was well-tolerated. One mother of a child with SMA type I thought that the use of the MI-E made her child worse after experiencing two hospitalizations for atelectasis in the month following the initiation its use. One year after the discontinuation of use, however, MI-E therapy was restarted successfully in this 22-month-old child to treat an acute episode of atelectasis, with almost complete resolution of volume loss in 1 day (Fig 2). For this child, adjustment of the pressure settings from those used in the initial trial resulted in the continuation of use of the device at home after hospital discharge to maintain effective airway clearance.

### Table 2—Pulmonary Function Test Results*

<table>
<thead>
<tr>
<th>Tests</th>
<th>FVC, % predicted</th>
<th>FEV1, % predicted</th>
<th>FEF25–75, % predicted</th>
<th>Pimax, cm H2O</th>
<th>Pemax, cm H2O</th>
</tr>
</thead>
<tbody>
<tr>
<td>DMD</td>
<td>10.6 ± 6.3†</td>
<td>11.6 ± 7.6</td>
<td>26.7 ± 22.2</td>
<td>22.4 ± 12.0</td>
<td>21.1 ± 3.0</td>
</tr>
<tr>
<td>MYO</td>
<td>22.1 ± 11.2</td>
<td>21.0 ± 8.5</td>
<td>21.7 ± 10.5</td>
<td>22.7 ± 17.3</td>
<td>17.6 ± 7.4</td>
</tr>
<tr>
<td>NS-NMD</td>
<td>36.3 ± 17.8</td>
<td>36.8 ± 19.3</td>
<td>45.2 ± 32.5</td>
<td>23.5 ± 14.4</td>
<td>10.4 ± 2.4</td>
</tr>
<tr>
<td>SMA type II</td>
<td>24.7 ± 10.1</td>
<td>25.6 ± 11.4</td>
<td>41.2 ± 18.7</td>
<td>41.4 ± 17.4†</td>
<td>15.5 ± 5.5</td>
</tr>
</tbody>
</table>

*Values given as mean ± SD. FEF25–75 = forced expiratory flow, midexpiratory phase.
†p < 0.05, compared with NMD and SMA.
‡p < 0.05, compared with all other groups.

**Figure 2.** *Left, A:* chest radiograph of 22-month-old girl with SMA type I and right upper lobe density. *Right, B:* 24 h later, following use of the MI-E and therapy with antibiotics, there is marked clearing of the right upper lobe density.
Two patients (3%) chose to use other devices after the initial trial, and three patients (5%) thought that the device was ineffective or unpleasant. The two patients who chose other devices were adolescents, both with SMA type II. One patient used the device by tracheostomy and thought that it contributed to her chronic abdominal pain. The other used the MI-E by facemask and complained of chest discomfort during the MI-E maneuvers. Three of these five patients utilized noninvasive positive-pressure ventilation. Two of the three patients who discontinued MI-E use due to unpleasantness were infants. Their parents interpreted their crying and agitation as a response to the treatment, which prompted its discontinuation. The remaining patient was a young adult with a tracheostomy, who thought that MI-E therapy made no difference in his airway clearance.

Mechanical insufflation/exsufflation was considered to be safe for the majority of patients. There were no episodes of pneumothorax or pulmonary hemorrhage, or need for emergency tracheostomy tube changes due to mucus plugging. One patient with DMD and acute respiratory failure experienced an abnormal cardiac rhythm (premature ventricular contractions) on the initial use of the MI-E device. He later used the device without difficulty, after resolution of his acute respiratory failure.

There were no episodes of symptomatic reflux noted by any of the patients or caregivers. For the group, 32 patients (52%) had previously been prescribed medication for gastroesophageal reflux on a daily basis, and 3 of those patients (9%) also had experienced a previous Nissen fundoplication.

Since the chart review, four patients have died of complications related to their disease or of disease progression, but without any correlation to use of the MI-E device. One patient with SMA type I was not being observed and aspirated a meal, a patient with DMD died of cardiomyopathy, and a toddler with NS-NMD died from nonrespiratory causes. One child died of her disease before the device was delivered to her home.

In general, MI-E use was viewed as effective by caregivers. The number of acute lower respiratory infections in this group of patients was small over the observation period and did not permit meaningful comparison with a pretreatment period. Four patients (6%) experienced improvement in chronic atelectasis after the institution of MI-E. In another five children (8%), the families noted a reduction in the frequency of pneumonia after beginning MI-E use. For all patients who have tolerated it, the use of the MI-E device has continued at a minimum of once daily to maintain comfort with the interface and to maintain the technique of using the device. During periods of respiratory illness, 22 families (39% of those who continued use of the device) were instructed to increase the pressure settings so as to remove secretions more effectively and to promote a better sense of well-being in the patient. Of the remaining 35 patients who continued to use the MI-E device, 22 did not require a change in the pressure prescription to clear their airway effectively, and 13 did not contract an illness severe enough to require a consult with our office regarding changes to the treatment plan. In addition to using increased pressures, increased frequency of use also occurred, with intervals of 1 to 2 h becoming the “routine” during periods of respiratory illness. All caregivers were instructed to use the MI-E device as often as required to reflect the need to cough more frequently during a respiratory illness.

**Discussion**

In this population of children with NMD, the use of the MI-E was generally safe, well-tolerated, and effective. The use of the device has gained widespread acceptance in our institution. Families for whom it was prescribed were taught the proper use of the device, and how to increase MI-E effectiveness during acute respiratory illnesses by changing pressures and the frequency of sessions.

Reported complications of MI-E use in adults are rare. These include nausea, bradycardia, tachycardia, and abdominal distention.

In our series, only one patient experienced a cardiac rhythm disturbance, and no patients reported complaints of abdominal distension. In contrast, the rapid decrease in intrathoracic pressure below the intraabdominal pressure associated with negative pressure exsufflation could potentially predispose the patient to gastroesophageal reflux with aspiration. Peak intragastric pressures with the MI-E, however, have been reported to be less than or equal to one third of those generated by unassisted coughing in healthy subjects. In our review, no episodes of aspiration or reflux were noted by any caregivers after the use of the MI-E. Similar experiences were reported by Bach in a largely adult population.

Previous research has demonstrated that cough peak flows (CPFs) relate directly to the ability to clear secretions from the respiratory tract. The normal CPF in adults exceeds 360 L/min. In an earlier study, Barach measured the expiratory flows of healthy subjects and compared them to those using an exsufflation device, and found that the mechanical device produced flows at 60% of spontaneous coughs, a value that is capable of clearing secretions from the airway. Airway clearance in adults is compromised at CPFs of < 160 L/min. Furthermore,
baseline CPF values of > 160 L/min also may be insufficient for patients with NMD during a respiratory infection, as these patients can experience decreased respiratory muscle function during respiratory illnesses. The most notable alteration in pulmonary function has been found to be P\textsubscript{efmax}, which was correlated to “cough capacity” in DMD patients.

In our group of study patients, we used P\textsubscript{efmax} when possible to guide our decision to initiate MI-E therapy. It was recommended that those patients who had experienced a decrease in their P\textsubscript{efmax} to < 60 cm H\textsubscript{2}O, or those who had an acute lower respiratory tract infection or atelectasis unresolved by conventional means, begin MI-E therapy. The use of lung function measurements to determine the need for airway clearance assistance may be problematic in children. Spirometry and measures of respiratory muscle strength are generally not performed in children < 5 to 6 years of age because of their inability to perform the tests reproducibly. Other patients with NMD and cognitive impairment, who would benefit from assistance with airway clearance, also cannot cooperate in following the instructions for pulmonary function test maneuvers. In addition, patients with NMD may not be referred for pulmonary evaluation and intervention before they have developed symptoms and have moderate-to-severe restrictive lung disease. Therefore, other methods or objective measurements have to be established to aid in identifying patients who are at risk for ineffective airway clearance.

We defined the effectiveness of the MI-E by patient report of secretion clearance or radiographic resolution of pneumonia or atelectasis. We did not routinely measure the amount of sputum recovered during the use of the MI-E. This would be difficult since many patients swallow secretions rather than expectorate them, and the majority of our patients did not have a tracheostomy tube to allow for easy secretion removal from the airway. Therefore, secretion clearance was evaluated subjectively. Patients often reported that it felt “easier to breathe” after the use of the MI-E, and that they could feel secretions being mobilized.

Intolerance of the MI-E in our patients may have been related to several factors. The most common reason for children in this series to oppose the use of the device was inadequate caregiver training regarding technique (e.g., asynchronous timing of breaths or using incorrect pressures). The reeducation of caregivers by hospital respiratory therapists or by the home equipment company therapists improved caregiver performance and patient acceptance. Parents of young children perceived intolerance if agitation occurred and/or if oxygen saturation decreased. These are often signs of airway obstruction after the dislodging of mucus with movement into proximal, larger airways. Teaching caregivers to anticipate this and how to resolve the symptoms promptly encouraged caregivers that the device could be tolerated and safe.

A limitation of this study is its retrospective nature. Our review concentrated on the characteristics of the patients who used MI-E therapy and was not a prospective study documenting changes in medical treatment as a result of MI-E therapy. A comparison between a pretreatment period and the time after MI-E use would not be practical, since many of the underlying NMDs included in our patient population are progressive. Thus, during the pretreatment period, patients would be expected to have better respiratory muscle and lung function, and fewer lower respiratory tract complications and hospitalizations, even without any specific airway clearance regimen. Thus, the apparent effectiveness of any intervention over a second period, after a decline of respiratory muscle strength and lung function have occurred, would be blunted when compared with pretreatment outcomes. There are, however, studies demonstrating the effectiveness of MI-E therapy for patients with DMD and SMA. These studies have recorded outcomes relative to hospitalizations, frequency of pneumonias, respiratory failure, and tracheotomy requirement.

Based on the premise that effective airway clearance prevents the complications associated with retained secretions, the use of the MI-E in pediatric patients with impaired cough may have several other positive benefits. These include a decreased need for antibiotics and supplemental oxygen, and a decreased number or duration of hospitalizations for respiratory illness. An increase in P\textsubscript{fmax} and FVC can result from improved elastic recoil that occurs with the reintroduction of higher lung volumes with positive insufflation.

In addition to the medical benefits of enhanced airway clearance, the importance of the psychological benefits to patients with NMD should be considered as well. An improvement in perceived quality of life due to fewer acute “illness-related” activities would help to promote the more long-term goals of “normal” or socially acceptable adult behavior. Until recently, many patients with NMD and their families have lived with the belief that their disease is terminal at a young age. Since medical intervention has been able to prolong life in people with NMD, the goals and activities of daily living associated with adulthood are possible. Quality-of-life studies involving the population affected by NMD have not been published. Further research is necessary to study the
relationship between the new medical therapies being used and the patient’s perspective on their life with the new therapies.

In summary, 90% of the patients with NMD evaluated for use of the MI-E in our program continue to utilize this therapy. No life-threatening complications or events requiring acute hospitalization occurred related to use of the device. Rapid resolution of atelectasis or decrease in frequency of pneumonia was documented in nine patients. Therefore, we conclude that MI-E use is safe, effective, and well-tolerated in a pediatric population.

REFERENCES

7 Bach JR. Mechanical insufflation-exsufflation: comparison of peak expiratory flows with manually assisted and unassisted coughing techniques. Chest 1993; 104:1553-1562
10 Segal MS, Salomon A, Herschfus JA. Alternating positive-negative pressures in mechanical respiration (the cycling valve device employing air pressures). Dis Chest 1954; 25:640-648
12 Barach AL, Beck GJ. Exsufflation with negative pressure: physiologic and clinical studies in poliomyelitis, bronchial asthma, pulmonary emphysema and bronchiectasis. Arch Intern Med 1954; 93:825-841
22 Bach JR. Continuous noninvasive ventilation for patients with neuromuscular disease and spinal cord injury. Semin Respir Crit Care Med 2002; 23:283-292