Dealing with airway clearance and daytime respiratory failure in children with neuromuscular disease

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Introduction

• Impaired cough strength is one of the first respiratory manifestations of progressive NMD

- “Air drives mucus”

- Efficient cough:
  1. Inspiration to TLC
  2. Close the glottis – short pause
  3. Efficient contraction of the expiratory muscles.
Introduction
Introduction

- Impaired evacuation of secretions:
  - NMD: no (initial) impaired mucociliary clearance
  - Daily production of mucus

- Respiratory tract infection:
  - Increased number of secretions
  - Purulent secretions
  - Impaired airway clearance
  - Decreased muscle strength
  - Development of atelectasis and respiratory insufficiency
Neurological and Neuromuscular Disease as a Risk Factor for Respiratory Failure in Children Hospitalized With Influenza Infection

**Figure.** Predicted Probability of Respiratory Failure in Children With Chronic Medical Conditions Hospitalized With Influenza Infection

NNMD indicates neurological and neuromuscular disease. None of the patients had all 3 chronic medical conditions. Error bars indicate 95% confidence intervals.
Effect of Upper Respiratory Tract Infection in Patients with Neuromuscular Disease

JANET M. POPONICK, I. JACOBS, GERALD SUPINSKI, and ANTHONY F. DI MARCO

Figure 1. Effects of acute URI on vital capacity, maximal inspiratory and expiratory pressures, transcutaneous oxygen saturation, and end-tidal PCO₂ in one subject with neuromuscular disease. See text for further explanation.
Effect of Upper Respiratory Tract Infection in Patients with Neuromuscular Disease

JANET M. POPONICK, I. JACOBS, GERALD SUPINSKI, and ANTHONY F. DiMARCO

### TABLE 3

<table>
<thead>
<tr>
<th></th>
<th>VC † (L)</th>
<th>MIP ‡ (cm H₂O)</th>
<th>MEP ‡ (cm H₂O)</th>
<th>PCO₂ (mm Hg)</th>
<th>SO₂ (mm Hg)</th>
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</thead>
<tbody>
<tr>
<td><strong>Baseline</strong></td>
<td>1.16 ± 0.14</td>
<td>49.2 ± 6.8</td>
<td>35.5 ± 3.8</td>
<td>39.1 ± 1.1</td>
<td>95.1 ± 1.0</td>
</tr>
<tr>
<td>(50.1% ± 6.6)</td>
<td>(51.5% ± 8.2)</td>
<td>(26.8% ± 3.9)</td>
<td></td>
<td></td>
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<tr>
<td><strong>URI</strong></td>
<td>1.01 ± 0.15 †</td>
<td>37.1 ± 6.2*</td>
<td>25.5 ± 3.0*</td>
<td>43.9 ± 2.1†</td>
<td>95.0 ± 1.0</td>
</tr>
<tr>
<td>(43.4% ± 6.5)</td>
<td>(38.9% ± 7.1)</td>
<td>(19.7% ± 2.8)</td>
<td></td>
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<tr>
<td><strong>Recovery</strong></td>
<td>1.09 ± 0.14</td>
<td>46.2 ± 7.0</td>
<td>34.1 ± 3.2</td>
<td>38.9 ± 1.2</td>
<td>95.3 ± 1.0</td>
</tr>
<tr>
<td>(48.5% ± 6.5)</td>
<td>(47.9% ± 8.4)</td>
<td>(25.8% ± 3.3)</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

* Significantly less than baseline values (p < 0.05).
† Significantly greater than baseline values (p < 0.05).
‡ Mean absolute and percent predicted values are provided.
Introduction

URTI
Introduction

Increased risk of LRTI:

- Multifactorial
- Hypoventilation
- Ineffective cough
- Scoliosis
- High prevalence of gastro-esophageal reflux and recurrent aspiration
- Nutritional status
Hospital Readmissions for Newly Discharged Pediatric Home Mechanical Ventilation Patients

Sheila S. Kun, RN, MS,1† Jeffrey D. Edwards, MD, MA,2*† Sally L. Davidson Ward, MD,1 and Thomas G. Keens, MD1

Justed odds ratios of select predictor variables for readmission. Results: The 12-month incidence of non-elective readmission was 40% Close to half of these readmissions occurred

<table>
<thead>
<tr>
<th>Reason</th>
<th>n (%)</th>
</tr>
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<tbody>
<tr>
<td>Pneumonia</td>
<td>22 (28)</td>
</tr>
<tr>
<td>Tracheitis</td>
<td>13 (17)</td>
</tr>
<tr>
<td>Tracheostomy decannulation/obstruction</td>
<td>9 (11.5)</td>
</tr>
<tr>
<td>Abdominal pain/emesis</td>
<td>8 (10)</td>
</tr>
<tr>
<td>Infectious, other</td>
<td>5 (6.5)</td>
</tr>
<tr>
<td>Gastrointestinal, other</td>
<td>4 (5)</td>
</tr>
<tr>
<td>Tracheostomy bleeding</td>
<td>4 (5)</td>
</tr>
<tr>
<td>Failure to thrive/feeding intolerance</td>
<td>3 (4)</td>
</tr>
<tr>
<td>Neurosurgical</td>
<td>3 (4)</td>
</tr>
<tr>
<td>Dehydration/electrolyte imbalance</td>
<td>2 (2.5)</td>
</tr>
<tr>
<td>Respiratory, other</td>
<td>2 (2.5)</td>
</tr>
<tr>
<td>Seizures</td>
<td>2 (2.5)</td>
</tr>
<tr>
<td>Ophthalmologic</td>
<td>1 (1.5)</td>
</tr>
<tr>
<td>Total</td>
<td>78</td>
</tr>
</tbody>
</table>
Introduction

Stage I  Intermittent hypoxemia and hypercapnia  →  REM sleep
Stage II  Nocturnal hypoventilation  →  REM + NREM sleep
Stage III  Hypoventilation during sleep and wakefulness  →  REM + NREM sleep + wakefulness
EVALUATION
Evaluation

- **History:**
  - Weak cough – sometimes only during infections
  - Prolonged cough after RTI
  - Recurrent RTI
Evaluation

- Lung function: VC, MIP, MEP, SNIP, cough peak flow:
  - Cut-off values have been proposed for respiratory failure and impaired cough strength.
  - Cough peak flow:
    - < 270 l/min: increased risk of atelectasis and respiratory failure
    - < 160 l/min: significantly impaired cough strength – predictive for extubation failure
      - Cough Peak Flows: Standard Values for Children and Adolescents. Bianchi C. et al., 2008: 50th percentiles were from 147 to 488 l/min and from 162 to 728 l/min in females and males, respectively, through an age range of 4–18 yrs, with levels in males being generally higher than those in females at any particular age.

- Polysomnography/polygraphy/capnography/blood gases
Predictors of severe chest infections in pediatric neuromuscular disorders

C. Dohna-Schwake \textsuperscript{a,*}, R. Ragette \textsuperscript{b}, H. Teschler \textsuperscript{b}, T. Voit \textsuperscript{a}, U. Mellies \textsuperscript{a}

Table 2

<table>
<thead>
<tr>
<th>Severe chest infections</th>
<th>IVC \leq 1.1 l (%)</th>
<th>IVC \leq 30% pred. (%)</th>
<th>PCF \leq 160 l/m (%)</th>
<th>PEP \leq 3 kPa (%)</th>
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</thead>
<tbody>
<tr>
<td>Sensitivity</td>
<td>90.5</td>
<td>61.9</td>
<td>75.2</td>
<td>85.7</td>
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<tr>
<td>Specificity</td>
<td>70.8</td>
<td>79.2</td>
<td>79.2</td>
<td>47.3</td>
</tr>
<tr>
<td>Ppv</td>
<td>73.1</td>
<td>63.2</td>
<td>79.2</td>
<td>62.1</td>
</tr>
<tr>
<td>Npv</td>
<td>89.5</td>
<td>65.4</td>
<td>76.2</td>
<td>76.9</td>
</tr>
</tbody>
</table>

IVC, inspiratory vital capacity; IVC pred., inspiratory vital capacity of predicted; PCF, peak cough flow; PEP, peak expiratory pressure; Ppv, positive predictive value; Npv, negative predictive value.
TREATMENT – AIRWAY CLEARANCE
Airway clearance techniques in neuromuscular disorders: A state of the art review

Michelle Chatwin\textsuperscript{a,*}, Michel Toussaint\textsuperscript{b}, Miguel R. Gonçalves\textsuperscript{c}, Nicole Sheers\textsuperscript{d}, Uwe Mellies\textsuperscript{e}, Jesus Gonzales-Bermejo\textsuperscript{f}, Jesus Sancho\textsuperscript{g}, Brigitte Fauroux\textsuperscript{h}, Tiina Andersen\textsuperscript{i}, Brit Hov\textsuperscript{j}, Malin Nygren-Bonnier\textsuperscript{k}, Matthieu Lacombe\textsuperscript{l}, Kurt Pernet\textsuperscript{b}, Mike Kampelmacher\textsuperscript{m}, Christian Devaux\textsuperscript{n}, Kathy Kinnett\textsuperscript{o}, Daniel Sheehan\textsuperscript{p}, Fabrizio Rao\textsuperscript{q}, Marcello Villanova\textsuperscript{r}, David Berlowitz\textsuperscript{d}, Brenda M. Morrow\textsuperscript{s}

\textit{Respiratory Medicine} 136 (2018) 98–110
Treatment – Cough Augmentation

- 2 components:
  1. Increase the inspiratory volume
  2. Augment the expiratory force
Treatment – Cough Augmentation

• Air-stacking:
  - Maximal insufflation capacity
  - Depending on function of bulbar muscles
  - Glossopharyngeal breathing
  - Bag and mask
  - Ventilator
  - Mechanical in- and exsufflator
One cycle of GI

Step 1: Air is drawn in through the mouth.

Step 2: The lips close and the air is trapped and pushed back with the tongue.

Step 3: The vocal cords open and the air is forced into the lungs.

Step 4: The vocal cords close and the air is trapped in the lungs.

After repeated cycles the air is exhaled passively.
Fig. 3. Improvement of peak cough flows in 27/29 pediatric patients with various muscle disorders. PCF0, PCF at spontaneous breathing; PCF1, PCF after IPPB hyperinsufflation.
Glossopharyngeal pistonning for lung insufflation in children with spinal muscular atrophy type II

Malin Nygren-Bonnier (malin.nygren-bonnier@karolinska.se)¹, Agneta Markström², Peter Lindholm³, Eva Mattsson¹, Brita Klefbeck¹

<table>
<thead>
<tr>
<th>Case</th>
<th>IVC pre (L)</th>
<th>IVC post (L)</th>
<th>PEF pre (L/min)</th>
<th>PEF post (L/min)</th>
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<tbody>
<tr>
<td>1</td>
<td>1.57</td>
<td>1.76</td>
<td>200</td>
<td>263</td>
</tr>
<tr>
<td>2</td>
<td>0.77</td>
<td>0.94</td>
<td>159</td>
<td>178</td>
</tr>
<tr>
<td>3</td>
<td>0.97</td>
<td>0.91</td>
<td>131</td>
<td>145</td>
</tr>
<tr>
<td>4</td>
<td>0.46</td>
<td>0.73</td>
<td>95</td>
<td>134</td>
</tr>
</tbody>
</table>

Pre = value before training; post = value after training. Mean change for IVC (0.13, 95% confidence interval 0.03–0.23) and PEF (116, 95% confidence interval 60–173).
Treatment – Cough Augmentation

- Augment expiratory force:
  - Manually assisted cough
  - Mechanical in- and exsufflation.
  - Combination
Fig. 1. Manually assisted cough via thoracic compression.

Fig. 2. Manually assisted cough via abdominal-thoracic compression.
Limits of Effective Cough-Augmentation Techniques in Patients With Neuromuscular Disease

Michel Toussaint PT PhD, Louis J Boitano MSc RRT, Vincent Gathot MSc PT, Marc Steens MSc PT, and Philippe Soudon MD

![Box plot showing peak cough flow for different techniques.](image-url)
Limits of Effective Cough-Augmentation Techniques in Patients With Neuromuscular Disease

Michel Toussaint PT PhD, Louis J Boitano MSc RRT, Vincent Gathot MSc PT, Marc Steens MSc PT, and Philippe Soudon MD
Treatment – Cough Augmentation

- Mechanical in- and exsufflator (Cough Assist):
  - Fauroux et al., Chest, 2008; settings:
    - Cough Assist.
    - 3 MI-E sessions with 6 cycli.
    - Pressures were increased from +/- 15 to +/- 40 cm H2O.
    - Insufflation of 2 sec. and exsufflation of 3 sec.
    - 30 sec rest between each sequence
    - 17 patients with a mean age of 11 years.
Treatment – Cough Augmentation
Treatment – Cough Augmentation

Best PEF or PCF (L/min)
Cough augmentation with mechanical insufflation/exsufflation in patients with neuromuscular weakness

M. Chatwin*, #, E. Ross#, N. Hart#, A.H. Nickol#, M.I. Polkey*, #, A.K. Simonds*

![Graph a) and b)](image-url)
Cough augmentation with mechanical insufflation/exsufflation in patients with neuromuscular weakness

M. Chatwin*,#, E. Ross#, N. Hart#, A.H. Nickol#, M.I. Polkey*,#, A.K. Simonds*

A further notable observation in the present study was that, during titration of mechanical insufflation/exsufflation pressures to patient comfort, the highest insufflation and exsufflation pressures did not necessarily produce the greatest PCF. One reason for this discrepancy could be vocal cord dysfunction, either due to the disease (despite exclusion of patients with moderate or severe bulbar involvement) or as a result of upper airway collapse, secondary to the application of positive or negative pressure during insufflation or exsufflation, respectively [26].
The Addition of Mechanical Insufflation/Exsufflation Shortens Airway-Clearance Sessions in Neuromuscular Patients With Chest Infection

Michelle Chatwin PhD and Anita K Simonds MD

Fig. 1. Mean ± SD treatment time for physiotherapy plus non-invasive ventilation (NIV) without insufflation/exsufflation (in-exsufflation) and with in-exsufflation, prior to the first assessment (white bars). The black bars show the mean and SD treatment time after the initial assessment at 30 min.
The Addition of Mechanical Insufflation/Exsufflation Shortens Airway-Clearance Sessions in Neuromuscular Patients With Chest Infection

Michelle Chatwin PhD and Anita K Simonds MD

Fig. 2. Visual-analog-scale scores (mean and SD) before the intervention (white bars) and at first assessment (30 min or earlier if chest clear) (black bars). The lower the score the more favorable the outcome. There was a significant decline in the reported amount of secretions for both interventions. However, there was a significantly higher level of fatigue with the intervention (in-exsufflation).
Airway clearance techniques in neuromuscular disorders: A state of the art review

Michelle Chatwin\textsuperscript{a,*}, Michel Toussaint\textsuperscript{b}, Miguel R. Gonçalves\textsuperscript{c}, Nicole Sheers\textsuperscript{d}, Uwe Mellies\textsuperscript{e}, Jesus Gonzales-Bermejo\textsuperscript{f}, Jesus Sancho\textsuperscript{g}, Brigitte Fauroux\textsuperscript{h}, Tiina Andersen\textsuperscript{i}, Brit Hov\textsuperscript{j}, Malin Nygren-Bonnier\textsuperscript{k}, Matthieu Lacombe\textsuperscript{l}, Kurt Pernet\textsuperscript{b}, Mike Kampelmacher\textsuperscript{m}, Christian Devaux\textsuperscript{n}, Kathy Kinnett\textsuperscript{o}, Daniel Sheehan\textsuperscript{p}, Fabrizio Rao\textsuperscript{q}, Marcello Villanova\textsuperscript{r}, David Berlowitz\textsuperscript{d}, Brenda M. Morrow\textsuperscript{s}

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Box 2
Recommendations For The Use Of Mechanical Insufflation-Exsufflation (MI-E).

- MI-E is the treatment of choice for the weaker group of patients with NMD
- Face masks should be used when using MI-E in patients without an artificial airway
- Inspiratory and expiratory timing/pressures should be individualized with progressive build-up of pressure until efficacy is achieved
- Higher expiratory than inspiratory pressures are advisable
- Patients with ALS are likely to benefit from lower pressures, triggered insufflation and longer insufflation time
- MI-E is possible through tracheostomy tubes, with higher pressures for smaller tube diameters
- Complete the session with an insufflation to leave an appropriate functional residual capacity in weaker patients or children
- In ICU, MI-E maybe as a useful technique to prevent re-intubation
- MI-E may be considered in the weaker children with bulbar insufficiency, and those who cannot cooperate with MAC or AS or in whom these methods are not effective
British Thoracic Society guideline for respiratory management of children with neuromuscular weakness

**Airway clearance and respiratory muscle training**

- Children with ineffective cough (including children over 12 years of age with cough peak flow <270 litres/min), particularly if they have had episodes of deterioration with respiratory infection, should be taught augmented cough techniques. [C]

- Manual cough assist and air-stacking methods to achieve maximum insufflation capacity are effective methods of improving cough efficiency and should be used when appropriate. [C]

- Mechanical insufflation/exsufflation (MI-E) should be considered in very weak children, those with loss of bulbar function, and those who cannot cooperate with manual cough assist or air-stacking or in whom these methods are not effective. [C]
Intrapulmonary Percussive Ventilation vs Incentive Spirometry for Children With Neuromuscular Disease

Christine Campbell Reardon, MD; Demian Christiansen; Elizabeth D. Barnett; Howard J. Cabral, PhD

Table 3. Clinical Outcomes for 18 Patients With NMD Treated With IS or IPV

<table>
<thead>
<tr>
<th></th>
<th>IS (n = 9)</th>
<th>IPV (n = 9)</th>
<th>IRR (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Days on antibiotics*</td>
<td>44</td>
<td>0</td>
<td>43 (6-333)</td>
</tr>
<tr>
<td>No. of pulmonary infections*</td>
<td>3</td>
<td>0</td>
<td>3.9 (.43-35)</td>
</tr>
<tr>
<td>School days missed for respiratory illness</td>
<td>5</td>
<td>0</td>
<td>4.8 (.5-37)</td>
</tr>
<tr>
<td>Days hospitalized for pulmonary function*</td>
<td>8</td>
<td>0</td>
<td>8.5 (1.1-67)</td>
</tr>
</tbody>
</table>
Effect of Intrapulmonary Percussive Ventilation on Mucus Clearance in Duchenne Muscular Dystrophy Patients: A Preliminary Report

Michel Toussaint PT, Harry De Win PT, Mark Steens PT, and Philippe Soudon MD

![Graph showing weight (g) comparison between With IPV and Without IPV conditions at T0, T1 + T2, T0, and T1 + T2.]
Airway clearance techniques in neuromuscular disorders: A state of the art review

Michelle Chatwin\textsuperscript{a,*}, Michel Toussaint\textsuperscript{b}, Miguel R. Gonçalves\textsuperscript{c}, Nicole Sheers\textsuperscript{d}, Uwe Mellies\textsuperscript{e}, Jesus Gonzales-Bermejo\textsuperscript{f}, Jesus Sancho\textsuperscript{g}, Brigitte Fauroux\textsuperscript{h}, Tiina Andersen\textsuperscript{i}, Brit Hov\textsuperscript{j}, Malin Nygren-Bonnier\textsuperscript{k}, Matthieu Lacombe\textsuperscript{l}, Kurt Pernet\textsuperscript{b}, Mike Kampelmacher\textsuperscript{m}, Christian Devaux\textsuperscript{n}, Kathy Kinnett\textsuperscript{o}, Daniel Sheehan\textsuperscript{p}, Fabrizio Rao\textsuperscript{q}, Marcello Villanova\textsuperscript{r}, David Berlowitz\textsuperscript{d}, Brenda M. Morrow\textsuperscript{s}

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Box 3
Recommendations for Peripheral ACT’s.

- Peripheral ACT should be commenced before and after clearing any secretions from the upper airway with proximal ACT’s
- Peripheral ACT’s do not require physical or intellectual patient co-operation
- Peripheral ACT’s is possible in infants, children and adults, even in the presence of a tracheostomy and/or bulbar failure
- Deflation by CWS strapping, is promising and worth evaluating in a clinical trial
- MT should be considered as a treatment option
- In the ventilatory dependent patient, peripheral ACT should be used in combination with ventilator support

ACT: airway clearance technique; IPV: intrapulmonary percussive ventilation; HFCWO: high frequency chest wall oscillations; CWS: chest wall strapping; MT: manual techniques.
Oscillatory techniques such as high-frequency chest wall oscillation and intrapulmonary percussive ventilation should be considered in children who have difficulty mobilising secretions or who have persistent atelectasis, despite use of other airway clearance techniques. [D]

Airway clearance techniques should be used during respiratory infection when oxygen saturation levels fall below 95% while the child is breathing room air. If the techniques being used fail to result in an increase in oxygen saturation to 95% or above, different methods of airway clearance should be used. This may require attendance at hospital for treatment. [D]

MI-E should be available in the acute setting in all hospitals that treat neuromuscular patients as an alternative method of airway clearance with the purpose of preventing deterioration and the need for intubation and mechanical ventilation. [D]
The effect of intrapulmonary percussive ventilation in pediatric patients: A systematic review

Eline Lauwers PT, MSc | Kris Ides PT, PhD | Kim Van Hoorenbeeck MD, PhD | Stijn Verhulst MD, PhD

TABLE 2  Key points

IPV is suggested to be a safe technique in spontaneously and mechanically ventilated children.

IPV is considered to be effective for airway clearance and could be an adequate alternative to other ACTs for children with respiratory diseases in general.

IPV appears to be superior to other ACTs in children with atelectasis, neurological/neuromuscular disease and mild/moderate acute bronchiolitis.

Only a limited number of studies with small sample sizes investigated the effects of IPV in the pediatric population.

Besides larger clinical trials with sufficient quality and the use of adequate outcome measures, future research should focus on a wider range of pathologies, such as children with CF with acute exacerbation, non-CF bronchiectasis, severe bronchiolitis, bronchopulmonary dysplasia and primary ciliary dyskinesia.
Acute effects of intrapulmonary percussive ventilation in COPD patients assessed by using conventional outcome parameters and a novel computational fluid dynamics technique

<table>
<thead>
<tr>
<th></th>
<th>Pre Mean value</th>
<th>Pre SD</th>
<th>Post Mean value</th>
<th>Post SD</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>VC (L)</td>
<td>2.75</td>
<td>0.43</td>
<td>2.81</td>
<td>0.41</td>
<td>0.273</td>
</tr>
<tr>
<td>FEV₁ (%p)</td>
<td>39.49</td>
<td>23.62</td>
<td>40.27</td>
<td>23.23</td>
<td>0.225</td>
</tr>
<tr>
<td>FEV₁/VC (%)</td>
<td>34.00</td>
<td>13.44</td>
<td>33.80</td>
<td>13.75</td>
<td>0.564</td>
</tr>
<tr>
<td>TLC (%p)</td>
<td>169</td>
<td>62</td>
<td>168</td>
<td>57</td>
<td>1</td>
</tr>
<tr>
<td>MEF 25 (L/s)</td>
<td>0.13</td>
<td>0.05</td>
<td>0.13</td>
<td>0.05</td>
<td>0.18</td>
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<tr>
<td>MEF 50 (L/s)</td>
<td>0.34</td>
<td>0.23</td>
<td>0.35</td>
<td>0.27</td>
<td>0.785</td>
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<tr>
<td>RV (L)</td>
<td>4.86</td>
<td>2.19</td>
<td>4.61</td>
<td>2.34</td>
<td>0.686</td>
</tr>
<tr>
<td>DLCO (mmol/min/kPa)</td>
<td>2.99</td>
<td>1.49</td>
<td>3.25</td>
<td>1.76</td>
<td>0.138</td>
</tr>
<tr>
<td>DLCO/VA (mmol/min/kPa/L)</td>
<td>0.75</td>
<td>0.43</td>
<td>0.80</td>
<td>0.46</td>
<td>0.066</td>
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<tr>
<td>MIP (kPa)</td>
<td>6.88</td>
<td>0.61</td>
<td>6.84</td>
<td>1.02</td>
<td>1</td>
</tr>
<tr>
<td>MEP (kPa)</td>
<td>11.54</td>
<td>1.14</td>
<td>10.22</td>
<td>2.72</td>
<td>0.068</td>
</tr>
<tr>
<td>Borg Dyspnea</td>
<td>4</td>
<td>0.71</td>
<td>3.4</td>
<td>0.89</td>
<td>0.083</td>
</tr>
</tbody>
</table>
iRaw change baseline to post IPV [%]
Acute effects of intrapulmonary percussive ventilation in COPD patients assessed by using conventional outcome parameters and a novel computational fluid dynamics technique

<table>
<thead>
<tr>
<th>Patient 1</th>
<th>Patient 2</th>
<th>Pre</th>
<th>Patient 3</th>
<th>Patient 4</th>
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<table>
<thead>
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ACUTE RESPIRATORY FAILURE
Combined Noninvasive Ventilation and Mechanical In-Exsufflator in the Treatment of Pediatric Acute Neuromuscular Respiratory Failure

Tai-Heng Chen, MD,1,2 Jong-Hau Hsu, MD,1,3,4 Jiunn-Ren Wu, MD,1,3 Zen-Kong Dai, MD, PhD,1,3 I-Chen Chen, MD,1 Wen-Chen Liang, MD,1 San-Nan Yang, MD, PhD,1,4 and Yuh-Jyh Jong, MD, DMSci1,4,5,6*

<table>
<thead>
<tr>
<th>TABLE 2 — Baseline Characteristics Between Patients With Different Outcomes</th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Success</td>
<td>12</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>Failure</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>$P$-value</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cases</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age (y/o)</td>
<td>9.6 ± 7.3</td>
<td>6.0 ± 7.5</td>
<td>0.4</td>
</tr>
<tr>
<td>PRISM-III</td>
<td>8.4 ± 4.2</td>
<td>8.0 ± 3.4</td>
<td>0.9</td>
</tr>
<tr>
<td>HR (bpm)</td>
<td>141.1 ± 16.8</td>
<td>138.0 ± 15.3</td>
<td>0.8</td>
</tr>
<tr>
<td>RR (b/min)</td>
<td>30.3 ± 8.1</td>
<td>33.8 ± 11.1</td>
<td>0.5</td>
</tr>
<tr>
<td>MAP (mmHg)</td>
<td>85.1 ± 26.8</td>
<td>91.7 ± 10.2</td>
<td>0.6</td>
</tr>
<tr>
<td>pH</td>
<td>7.29 ± 0.07</td>
<td>7.20 ± 0.17</td>
<td>0.1</td>
</tr>
<tr>
<td>PaCO$_2$ (mmHg)</td>
<td>71.7 ± 18.6</td>
<td>77.6 ± 22.4</td>
<td>0.6</td>
</tr>
<tr>
<td>PaO$_2$ (mmHg)</td>
<td>98.2 ± 43.4</td>
<td>112.4 ± 40.7</td>
<td>0.6</td>
</tr>
<tr>
<td>PaO$_2$/FiO$_2$</td>
<td>307.9 ± 208.1</td>
<td>397.9 ± 156.2</td>
<td>0.4</td>
</tr>
<tr>
<td>SpO$_2$ (%)</td>
<td>97.0 ± 2.5</td>
<td>95.3 ± 5.3</td>
<td>0.4</td>
</tr>
</tbody>
</table>

PRISM-III, Pediatric Risk of Mortality-III score; HR, heart rate; RR, respiratory rate; MAP, mean arterial pressure.

$^1$Values are given as the mean ± SD, unless otherwise indicated.
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Fig. 1. Changes of physiologic indices over 24 hr after use of NIV/MIE in the success group. A: PaCO₂ level declined significantly as early as 12 hr after treatment. B: Acidosis improved after 24 hr of treatment. C: The PaO₂/FI₂O₂ ratio did not change significantly after treatment. * P < 0.001.
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To analyze factors associated with NIV failure, we compared physiologic parameters between groups and found that patients in the failure group had lower RR decrease from initial RR at 3 hr (success: 11.3 ± 8.9 vs. failure: 5.7 ± 7.6, $P = 0.02$). In addition, patients in the failure group had higher RR and HR at 9 hr when compared to those in the success group (HR: 139 ± 5.0 vs. 126.5 ± 10.8, $P = 0.04$; RR: 34.8 ± 5.4 vs. 28.2 ± 4.7, $P = 0.03$).
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All patients tolerated NIV and MIE well. Sedation by chloral hydrate or midazolam was required in four patients younger than 3 years old. The initial physical indices and outcomes were comparable between patients with and without sedation (data not shown). There was no major complication such as pneumothorax or gastric distension. The most common complication was skin irritation caused by NIV interfaces, including mild erosion or irritant dermatitis of nasal bridge in five cases. All these skin lesions healed completely with supportive care during the ICU stay and did not affect the use of NIV or MIE.
DAYTIME RESPIRATORY FAILURE
Daytime respiratory failure

- Characterised by daytime hypercapnia.
- Progressive decompensation – usually preceded by sleep-related hypoventilation and by recurrent episodes of respiratory exacerbations.

- Poor prognosis if left untreated.
- Association with low VC (25%)
Daytime respiratory failure

- Treatment:
  - Daytime NIV
  - Nasal mask/pillows
  - Mouthpiece ventilation
Daytime respiratory failure

- MPV:
  - Intermittent ventilation
  - Minimalizes facial pressure
  - Facilitates speech, eating and swallowing.
  - Better appearance
  - Decrease aerophagia
  - Dedicated ventilatory modes, interfaces and circuits with very sensitive triggers.
  - Some construction is often needed to allow access according to the patient’s needs.
CONCLUSIONS
Conclusions

• Impaired cough is an important factor in the management of patients with NMD

• Various approaches exist – a combination of techniques is often applied

• Treatment should be individualized

• Daytime respiratory failure: workshop on mouthpiece ventilation