Pro-Con Debate:
Mechanically Assisted Insufflation-Exsufflation (MIE) in Neuromuscular Disease

Aaron St-Laurent
Cross Canada Rounds, Jan. 2017
MIE Debate - Contents

• Cough primer
  – Purpose and physiology
  – Cough in neuromuscular disease (NMD)
  – Peak cough flow
  – Assisted cough & mechanical insufflation-exsufflation

• Pro-Side
  – Effect on peak cough
  – Effect on physiologic measures
  – Effect on outcomes
  – Safety and Acceptability

• Con-Side

• Recommendations

• Discussion
Cough

• Purpose: Airway clearance
  – Inhaled/aspirated materials
  – Retained secretions

• Voluntary vs. Involuntary
  – Cough receptors
    • Mechanical, Chemical irritant
    • Afferent – vagus
    • Efferent – phrenic, spinal motor nerves, vagus
Cough - Sequence

Phases of cough:

- Inspiratory
- Compressive
- Expulsive

Cough in NMD

• Inspiratory muscle weakness
  –Limits expiratory-muscle length tension relationship, chest wall recoil, decreased alveolar pressures

• Bulbar dysfunction
  –Effects compressive phase

• Expiratory muscle weakness
  –Decreased dynamic airway pressure, velocity
Cough in NMD

- Respiratory illness worsens weakness

What is peak cough flow?

- PCF a measure of cough effectiveness
- Measured via peak flow meter or spirometer
- Following complete inspiration, patient coughs forcibly
- Mouthpiece or face mask
- Normal adult peak cough >600L/min
Is Peak Cough Flow Important?

• Bach & Saporito:
  - Factors predicting successful extubation / decannulation in NM weakness
  - N=63, extubation/decannulation attempts
  - Studied: Age, ventilator use, FVC, PCF
  - PCF only predictor of success
    • All 43 attempts >160L/min – successful
    • All 15 attempts <160L/min – unsuccessful

• Bach et al.:
  - No patients with DMD and assisted peak cough >270L/min developed acute respiratory distress


Is Peak Cough Flow Important?

- Dohna-Schwake:
  - Retrospective
  - N=46, children with NMD without cough assist
    - Mean age 12.5 ± 3.6 years
  - Comparison
    - With pneumonia admission, N = 22
    - Without pneumonia admission, N = 26

Is Peak Cough Flow Important?

- PCF <160L/min – sensitive, specific predictor of severe chest infections
- PCF, IVC & FEV1 all significantly different between groups

Determinants of Peak Cough Flow

• Trebbia - Peak Cough Determinants in NMD
  – N=155,
  – Age 43 +/- 16 (16-80)
• Chief Determinants
  – Vital capacity
  – Mean inspiratory capacity
• Contribution to PCF Variance
  – MIC > ERV > MEP

Univariate regression analysis between PCF and other non-invasively measured variables

<table>
<thead>
<tr>
<th></th>
<th>PCF</th>
<th></th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>VC</td>
<td>0.58</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>MIC</td>
<td>0.47</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>MIP*</td>
<td>0.46</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>TLC</td>
<td>0.34</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>MEP*</td>
<td>0.31</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>ERV*</td>
<td>0.23</td>
<td></td>
<td>&lt;0.0001</td>
</tr>
<tr>
<td>FRC*</td>
<td>0.07</td>
<td></td>
<td>&lt;0.001</td>
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</tbody>
</table>

Stepwise regression analysis between PCF and other non-invasively measured variables (excluding VC and TLC)

<table>
<thead>
<tr>
<th></th>
<th>PCF</th>
<th></th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>MIC</td>
<td>0.44</td>
<td></td>
<td>0.0001</td>
</tr>
<tr>
<td>ERV*</td>
<td>0.13</td>
<td></td>
<td>0.0001</td>
</tr>
<tr>
<td>MEP*</td>
<td>0.02</td>
<td></td>
<td>0.0001</td>
</tr>
</tbody>
</table>

Airway Clearance Techniques

• Lung Volume Recruitment Manoeuvers
  – Glossopharyngeal breathing
  – Manual breath-stacking (ambubag)
  – Mechanical insufflation (BiPAP, MIE)

• Expiratory flow manoeuvres
  – Manually assisted cough
  – Mechanical exsufflation

• Combination
Mechanical Insufflation-Exsufflation

- First described 1954
- Manual vs. automatic timed
- Pressure-limited insufflation phase
- Rapid reversal to negative pressure
- Exsufflation phase to clear secretions
Mechanical Insufflation-Exsufflation
Pro-Side

Mechanical Insufflation-Exsufflation is an important airway clearance technique in patients with neuromuscular disease
Effect on Cough

• Bach: Comparison of PCF with various assisted cough methods (n=21, adults)
  – N=21, ventilator-dependent adults, VC=490mL

Unassisted PCF = 1.81 ± 1.03 L/s
MIC (breath stacking) = 3.37 ± 1.07 L/s
MIC & manual cough = 4.27 ± 1.29 L/s
Mechanical cough = 7.47 ± 1.02 L/S

• p<0.001

4) Bach et al; Mechanical insufflation-exsufflation. Comparison of peak expiratory flows with manually assisted and unassisted coughing techniques. Chest. 1993 Nov;104(5):1553-62
Effect on Cough

• Chatwin
  – Comparison of peak cough flow techniques
    • Physiotherapy, BiPAP, exsufflation, MIE
  – Pediatric N=8, Adult N=14.
  – Age matched controls
  – Ineffective cough or respiratory infections

Effect on Cough

Pediatric
N=8
Age 13.6 ± 2.4

Adults
N=12
Age 26 ± 8

** , ***: p=0.01, p=0.001 versus UAC.

Effect on Cough

MIE – Effect on Cough

- Faroux:
  - MIE at various pressures
  - 17 consecutive pediatric NMD outpatients
  - Stable at least 1-months

Table 1—Characteristics of the Patients*

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Gender</th>
<th>Age, yr</th>
<th>Weight, kg</th>
<th>Able to Walk, No.</th>
<th>NPPV, No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Duchenne muscular dystrophy (n = 4)</td>
<td>Boy</td>
<td>15 ± 3</td>
<td>36 ± 22</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td></td>
<td>Girl</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Spinal muscular atrophy (n = 4)</td>
<td>Boy</td>
<td>8 ± 3</td>
<td>16 ± 3</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td></td>
<td>Girl</td>
<td>2</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other neuromuscular disease (n = 9)</td>
<td>Boy</td>
<td>10 ± 2</td>
<td>38 ± 17</td>
<td>2</td>
<td>5</td>
</tr>
<tr>
<td></td>
<td>Girl</td>
<td>3</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total population (n = 17)</td>
<td>Boy</td>
<td>11 ± 4</td>
<td>33 ± 18</td>
<td>4</td>
<td>10</td>
</tr>
<tr>
<td></td>
<td>Girl</td>
<td>5</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*Values are given as the mean ± SD, unless otherwise indicated. NPPV = noninvasive positive pressure ventilation.

Effect on Cough

- Pressures ± 40
  - Greatest effect on VC, Inspiratory/Expiratory flows, and PCF
  - Tolerated

Effect on Physiologic Measures

Stehling: Retrospective, N=21, placed on MIE

- VC <30% predicted, PCF < 160L/min, nocturnal NIV
- 16 failed intermittent positive pressure breathing for assisted cough
- Home MIE, 10-minutes, twice daily

Table 1. Patient characteristics.

<table>
<thead>
<tr>
<th></th>
<th>Duchenne muscular dystrophy (n = 10)</th>
<th>Spinal muscular atrophy (n = 4)</th>
<th>Congenital myopathy without diaphragm weakness (n = 4)</th>
<th>Congenital myopathy with diaphragm weakness (n = 3)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (male/female)</td>
<td>10/0</td>
<td>3/1</td>
<td>2/2</td>
<td>2/1</td>
</tr>
<tr>
<td>Age (years)</td>
<td>20.6 ± 3.9 (15–27)</td>
<td>9.3 ± 1.7 (7–11)</td>
<td>14.0 ± 9.6 (5–27)</td>
<td>13.7 ± 0.6 (13–14)</td>
</tr>
<tr>
<td>VC (l)</td>
<td>0.48 ± 0.15 (0.25–0.68)</td>
<td>0.34 ± 0.13 (0.25–0.53)</td>
<td>0.54 ± 0.39 (0.33–1.12)</td>
<td>0.62 ± 0.32 (0.52–1.09)</td>
</tr>
<tr>
<td>VC (% predicted)</td>
<td>11.0 ± 3.5 (5–15)</td>
<td>15.0 ± 2.6 (12–18)</td>
<td>28.0 ± 20.4 (10–54)</td>
<td>23.3 ± 6.6 (19–31)</td>
</tr>
<tr>
<td>Noninvasive ventilation</td>
<td>10/10 (100%)</td>
<td>4/4 (100%)</td>
<td>4/4 (100%)</td>
<td>3/3 (100%)</td>
</tr>
<tr>
<td>Hyperinsufflation</td>
<td>7/10 (70%)</td>
<td>3/4 (75%)</td>
<td>3/4 (75%)</td>
<td>3/3 (100%)</td>
</tr>
</tbody>
</table>

Effect on Physiologic Measures

- Significant increase in VC (p=0.002)
- Vital capacity increased in 18/21
  - Average relative increase 28%
- Effect remained stable

Table 2. Course of VC before and after the initiation of mechanical insufflation/exsufflation.

<table>
<thead>
<tr>
<th></th>
<th>-2 years (n = 16)</th>
<th>-1 year (n = 14)</th>
<th>Start M-I/E (n = 21)</th>
<th>+1 year (n = 21)</th>
<th>+2 years (n = 6)</th>
</tr>
</thead>
<tbody>
<tr>
<td>VC (L)</td>
<td>0.88 ± 0.45</td>
<td>0.71 ± 0.38</td>
<td>0.50 ± 0.24</td>
<td>0.64 ± 0.28</td>
<td>0.65 ± 0.29</td>
</tr>
<tr>
<td></td>
<td>(0.22–1.62)</td>
<td>(0.25–1.49)</td>
<td>(0.25–1.12)</td>
<td>(0.24–1.40)</td>
<td>(0.28–1.15)</td>
</tr>
<tr>
<td>VC (% predicted)</td>
<td>29.3 ± 18.8 (4–86)</td>
<td>21.3 ± 12.1 (5–45)</td>
<td>16.7 ± 10.9 (5–54)</td>
<td>22.9 ± 16.8 (7–73)</td>
<td>20.0 ± 11.7 (7–39)</td>
</tr>
<tr>
<td>Time (months)</td>
<td>28.4 ± 5.5</td>
<td>13.4 ± 3.1</td>
<td>0</td>
<td>9.4 ± 2.8</td>
<td>25.7 ± 7.4</td>
</tr>
</tbody>
</table>

MIE - Effect on Outcomes: Short-Term

- Vianello: MIE in Consecutive patients with NMD in ICU with dyspnea and chest infection
  - N=11, 16 historical controls

<table>
<thead>
<tr>
<th>TABLE 2 Summary data at study entry</th>
<th>MIE</th>
<th>Control</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Group A</td>
<td>Group B</td>
</tr>
<tr>
<td>Number of subjects</td>
<td>11</td>
<td>16</td>
</tr>
<tr>
<td>Age, yrs</td>
<td>34.91 ± 17.28</td>
<td>39.75 ± 21.56</td>
</tr>
<tr>
<td>Body mass index</td>
<td>18.76 ± 3.57</td>
<td>20.32 ± 5.08</td>
</tr>
<tr>
<td>Sex (males, females)</td>
<td>7, 4</td>
<td>12, 4</td>
</tr>
<tr>
<td>Patients administered home NPPV, n</td>
<td>7</td>
<td>9</td>
</tr>
<tr>
<td>No. of hospitalizations in 1 year</td>
<td>0.64 ± 0.81</td>
<td>0.58 ± 0.67</td>
</tr>
<tr>
<td>FVC, liters</td>
<td>0.38 ± 0.08</td>
<td>0.97 ± 0.88</td>
</tr>
<tr>
<td>PEF, liters/sec</td>
<td>1.54 ± 0.74</td>
<td>2.12 ± 1.14</td>
</tr>
<tr>
<td>Gilardeau score</td>
<td>2.09 ± 1.70</td>
<td>1.38 ± 1.67</td>
</tr>
<tr>
<td>Patients with fever, n (temperature &gt; 38°C)</td>
<td>1</td>
<td>3</td>
</tr>
<tr>
<td>Patients with leukocytosis, n (WBC &gt; 12,000 × 10^9/liter)</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>PaO₂, mm Hg</td>
<td>61.5 ± 5.84</td>
<td>66.25 ± 10.48</td>
</tr>
<tr>
<td>PaCO₂, mm Hg</td>
<td>52.18 ± 12.06</td>
<td>48.25 ± 14.46</td>
</tr>
<tr>
<td>pH</td>
<td>7.36 ± 0.07</td>
<td>7.40 ± 0.07</td>
</tr>
</tbody>
</table>

NPPV, noninvasive positive-pressure ventilation; FVC, forced vital capacity; PEF, peak expiratory flow; WBC, white blood cell count.

Effect on Outcomes: Short-Term

Reduction in intubation and/or tracheostomy

Effect on Outcomes: Short-Term

- Miske: Retrospective, N=63, MIE users
  - Median use 13.4 mo
  - 4 (6%) – Resolution of chronic atelectasis
  - 5 (8%) – Decreased pneumonia frequency

### 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 ventilation</td>
<td>5</td>
<td>4</td>
<td>4</td>
<td>3</td>
<td>9</td>
</tr>
<tr>
<td>Tracheostomy with 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>
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</tbody>
</table>
*Values in parentheses are ranges.

### 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>Pmax, 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>41.1 ± 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.

Effect on Outcomes: Long-Term

Bach: Retrospective, protocol in DMD adults “high-risk”
- PCF <270L/min OR respiratory failure
- Protocol: IPPV, manual & mechanical cough during illness, sat monitor and access to the medical team

Table 1—Hospitalization Rate Comparisons for Nonprotocol Preventilator Use High-Risk Patients vs Protocol Part-time Noninvasive IPPV Users*

<table>
<thead>
<tr>
<th></th>
<th>Entire Population</th>
<th>Nonprotocol</th>
<th>Protocol</th>
<th>P Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>17</td>
<td>24</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalizations/pt</td>
<td>2.41±1.84</td>
<td>0.5±1.0</td>
<td>&lt;0.005</td>
<td></td>
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<tr>
<td>Incidence of hospitalizations/yr/pt</td>
<td>2.25±4.75</td>
<td>0.2±0.5</td>
<td>&lt;0.005</td>
<td></td>
</tr>
<tr>
<td>Hospitalizations avoided/pt</td>
<td>1.8±1.7</td>
<td>0.8±1.0</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalizations avoided/yr/pt</td>
<td>0.8±1.0</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalization days/pt</td>
<td>35.4±66.3</td>
<td>3.6±8.7</td>
<td>&lt;0.005</td>
<td></td>
</tr>
<tr>
<td>Average hospitalization days/yr/pt</td>
<td>21.4±37.8</td>
<td>1.8±5.2</td>
<td>&lt;0.005</td>
<td></td>
</tr>
<tr>
<td>Years</td>
<td>3.6±2.7</td>
<td>3.1±3.2</td>
<td>0.21</td>
<td></td>
</tr>
</tbody>
</table>

*This does not include the hospitalizations for introduction of definitive ventilator use. Pt=patient.

Effect on Outcomes: Long-Term

- Decreased hospitalization rate, days
- Tracheosomtomy vs. full-time non-invasive
  - Most tracheostomy patients “Full-Time”
  - Full-time >16hr/day
  - Difference access to MIE

Table 2—Hospitalization Rate Comparisons for Tracheostomy IPPV Users and Protocol Noninvasive IPPV Users*

<table>
<thead>
<tr>
<th></th>
<th>Nonprotocol</th>
<th>Part-time</th>
<th>Full-time</th>
<th>p Value¹</th>
<th>Noninvasive</th>
<th>Full-time</th>
<th>p Value¹</th>
</tr>
</thead>
<tbody>
<tr>
<td>Patients</td>
<td>22</td>
<td>24</td>
<td>14</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalizations</td>
<td>1.6±2.1</td>
<td>0.5±1.0</td>
<td>0.4±0.8</td>
<td>0.32</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalizations/yr/pt</td>
<td>0.3±0.4</td>
<td>0.2±0.5</td>
<td>0.1±0.4</td>
<td>0.34</td>
<td></td>
<td></td>
<td>0.04</td>
</tr>
<tr>
<td>Hospitalizations avoided/pt</td>
<td>1.8±1.7</td>
<td>1.3±1.4</td>
<td>1.1±5.2</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalizations avoided/yr/pt</td>
<td>0.8±1.1</td>
<td>0.5±0.8</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalization days/pt</td>
<td>17.2±22.1</td>
<td>3.6±8.7</td>
<td>4.5±3.6</td>
<td>0.01</td>
<td>0.01</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Hospitalization days/yr/pt</td>
<td>2.3±2.4</td>
<td>1.8±5.2</td>
<td>0.3±2.4</td>
<td>0.71</td>
<td>0.04</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Years</td>
<td>7.6±6.2</td>
<td>3.1±3.2</td>
<td></td>
<td></td>
<td>4.5±3.6</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*This does not include the hospitalizations for introduction of definitive ventilatory support. Pt=patient.
¹Comparison with nonprotocol patients.

Effect on Outcomes: Long-Term

Tzeng: Retrospective, protocol in NMD, adults & children
- PCF <270L/min OR respiratory failure
- Protocol: IPPV, manual & mechanical cough during illness, sat monitor and access to the medical team

<table>
<thead>
<tr>
<th>Category</th>
<th>Preprotocol</th>
<th>Protocol</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hospitalizations/yr/patient (n = 10), No.*</td>
<td>1.06 ± 0.84</td>
<td>0.03 ± 0.11</td>
<td>0.003</td>
</tr>
<tr>
<td>Hospitalization d/yr/patient (n = 10), No.*</td>
<td>20.76 ± 36.01</td>
<td>0.06 ± 0.20</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>yr/patient</td>
<td>3.42 ± 3.36</td>
<td>1.94 ± 0.74</td>
<td>0.26</td>
</tr>
<tr>
<td>Hospitalizations/yr/patient (n = 33), No.†</td>
<td>1.40 ± 1.96</td>
<td>0.08 ± 0.17</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>Hospitalization d/yr/patient (n = 33), No.†</td>
<td>20.14 ± 41.15</td>
<td>1.43 ± 3.71</td>
<td>&lt; 0.001</td>
</tr>
<tr>
<td>yr/patient</td>
<td>0.75 ± 1.48</td>
<td>0.09 ± 0.38</td>
<td>0.002</td>
</tr>
<tr>
<td>Intubations, No.</td>
<td>0.75 ± 1.48</td>
<td>0.09 ± 0.38</td>
<td>0.002</td>
</tr>
<tr>
<td>Days intubated (n = 12), No.‡</td>
<td>9.21 ± 17.06</td>
<td>2.85 ± 13.37</td>
<td>0.003</td>
</tr>
<tr>
<td>yr/patient</td>
<td>5.89 ± 6.89</td>
<td>3.91 ± 3.50</td>
<td>0.50</td>
</tr>
</tbody>
</table>

MIE - Tolerability & Acceptability

- Miske: Retrospective, pediatric, N=63
  - Median use 13.4 mo
- Well-tolerated (90%)
  - 2 (3%) Chose other devices
    - Contribution to chronic abdominal pain
    - Chest discomfort
  - 3 (5%) Ineffective and/or unpleasant
    - 2 infants – crying, agitation
    - Young adult – no effect on airway clearance

MIE - Adverse Events

- Adverse events = Rare
  - No pneumothorax in pediatric population
  - No pulmonary haemorrhage, barotrauma.

- Miske\textsuperscript{8}: N=63, pediatric
  - No symptomatic reflux
  - PVCs in one patient, first-use, acute illness
  - When used only during respiratory illness, caregivers were less comfortable and less effective with the device.

- Faroux\textsuperscript{6}: N=17, pediatric
  - No abdominal distension, reflux, chest pain

- Vianello\textsuperscript{7}: N=11, adult
  - 1 GER, 1 epistaxis, 1 abdominal distension
Summary

Mechanical insufflation-exsufflation in NMD

– Increased peak cough flow
– Increased VC with regular use
– Decreases need to intubation, tracheostomy
– Improvement in chronic atelectasis, and
decrease frequency of pneumonia
– Decreases need to hospitalization
– Safe and well-tolerated
Recommendations

British Thoracic Society guideline for respiratory management of children with neuromuscular weakness

Evidence statements
Use of airway clearance techniques which increase CPF, as part of a homecare treatment package which includes NIV, is associated with decreased hospital admission for respiratory infection and improved survival (evidence level 3). Several airway clearance techniques are available. The most effective method for a particular child depends on the ability of the child to cooperate with treatment and the severity of the child’s weakness (evidence level 3).

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]
Recommendations

- 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]

- Children who use regular night-time or diurnal NIV should use their ventilator to support deep breathing during airway clearance treatments. Use of NIV during airway clearance sessions can help prevent respiratory muscle fatigue. [✓]

- Children who use MI-E for airway clearance should be given long enough periods of rest during treatment sessions to prevent respiratory muscle fatigue due to coughing. [✓]

- At the end of a treatment session with MI-E it is important to complete the session with an insufflation to leave the child with an appropriate functional residual capacity. [✓]
Recommendations

American Thoracic Society Documents

Respiratory Care of the Patient with Duchenne Muscular Dystrophy
ATS Consensus Statement

Recommendations

• Patients with DMD should be taught strategies to improve airway clearance and how to employ those techniques early and aggressively.

• Use assisted cough technologies in patients whose clinical history suggests difficulty in airway clearance, or whose peak cough flow is less than 270 L/minute and/or whose maximal expiratory pressures are less than 60 cm H₂O.

• The committee strongly supports use of mechanical insufflation-exsufflation in patients with DMD and also recommends further studies of this modality.

• Home pulse oximetry is useful to monitor the effectiveness of airway clearance during respiratory illnesses and to identify patients with DMD needing hospitalization (8).
Recommendations

The Respiratory Management of Patients With Duchenne Muscular Dystrophy: A DMD Care Considerations Working Group Specialty Article

David J. Birnkrant, MD,1* Katharine M.D. Bushby,2 Raouf S. Amin, MD,3 John R. Bach, MD,4 Joshua O. Benditt, MD,5 Michelle Eagle, PhD,2 Jonathan D. Finder, MD,6 Maninder S. Kalra, MD,3 John T. Kissel, MD,7 Anastassios C. Koumbourlis, MD,8 and Richard M. Kravitz, MD9

Step 1: Volume Recruitment / Deep Lung Inflation Technique

- Volume recruitment / deep lung inflation technique (by self-inflating manual ventilation bag or mechanical in-/ex-sufflation) when FVC < 40% predicted

Step 2: Manual and Mechanically Assisted Cough Techniques

Necessary when:
- Respiratory infection present and baseline peak cough flow < 270 lpm*
- Baseline peak cough flow < 160 lpm or max expiratory pressure < 40cm water
- Baseline FVC < 40% predicted OR < 1.25 liters in older teen / adult

* All specified threshold values of peak cough flow and maximum expiratory pressure apply to older teenage and adult patients
For at-risk individuals and patients using NIV:
1. Education and preventive strategies in airway clearance must precede the need for mechanical ventilation whenever possible. (Consensus)
2. In the absence of contraindications, LVR techniques should be introduced with measurement of PCFs and MIC in those with PCFs<270 L/min. (GRADE 1C)
3. MAC is recommended alone or in addition to LVR to increase PCFs to >270 L/min. (GRADE 1C)
4. In the absence of contraindications, MI-E should be recommended for patients unable to achieve PCFs >270 L/min with LVR and/or MAC particularly during respiratory infection. (GRADE 1C)
Figure 1-2) Flow diagram for preventive airway clearance techniques. LVR: Lung volume recruitment; PCF: Peak cough flow. *Note: Manually Assisted Cough can be added to increase effectiveness of CoughAssist® (Philips Heathcare, USA)
• Trebbia: Cough determinants in NMD
• Boitano: Cough physiology
• Chatwin: Comparison of PCF using different methods
• Faroux: Tolerance, Physio benefits (CO2, Respiratory comfort)
• LT: Merino: Incr Survival in DMD with Mech Cough used during URI
• ST: Miske: Peds, Tolerance, Decreased atelecatasis, and PNA’
• Toussaint: Useless
• LT: Tzeng: Avoidance of hosp with IPPV and cough-assist
• ST: Vianello: MIE avoids Rx failure / ETT in NMD resp illness
• LT: Bach – DMD: Protocol of IPPV, Cough assist – avoid hosp in DMD
• Bach – Comparison of cough augmentation techniques
Effect on Cough

- Significant: PCF, Respiratory Comfort (VAS)

Table 3—Evolution of Breathing Pattern, Gas Exchange, and Respiratory Muscle Function After the MI-E Sessions With 15, 30, and 40 cm H₂O*

<table>
<thead>
<tr>
<th>Variables</th>
<th>Baseline</th>
<th>After MI-E</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>15 cm H₂O</td>
</tr>
<tr>
<td>Breathing pattern</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VT, L</td>
<td>0.27 ± 0.11</td>
<td>0.27 ± 0.13</td>
</tr>
<tr>
<td>fr, breaths/min</td>
<td>26 ± 11</td>
<td>27 ± 12</td>
</tr>
<tr>
<td>Ve, L/min</td>
<td>6.3 ± 1.9</td>
<td>5.9 ± 1.5</td>
</tr>
<tr>
<td>Gas exchange</td>
<td></td>
<td></td>
</tr>
<tr>
<td>SpO₂, %</td>
<td>97.1 ± 2.1</td>
<td>96.6 ± 2.6</td>
</tr>
<tr>
<td>PETCO₂, mm Hg</td>
<td>39.9 ± 3.8</td>
<td>38.0 ± 4.4</td>
</tr>
<tr>
<td>Respiratory muscle function</td>
<td></td>
<td></td>
</tr>
<tr>
<td>VC, L</td>
<td>1.04 ± 1.13</td>
<td>1.01 ± 1.05</td>
</tr>
<tr>
<td>SNIP, † cm H₂O</td>
<td>29 ± 19</td>
<td>30 ± 19</td>
</tr>
<tr>
<td>PEF or PCF, L/min</td>
<td>162 ± 97</td>
<td>173 ± 112</td>
</tr>
<tr>
<td>Respiratory comfort, VAS/100</td>
<td>73 ± 21</td>
<td>75 ± 18</td>
</tr>
</tbody>
</table>

*Values are given as the mean ± SD, unless otherwise indicated. NS = not significant.
†Analysis of variance.
‡Analysis of variance was performed in 16 patients.