Respiratory Management—Your Questions Answered!

Michelle Chatwin, PhD
Consultant Physiotherapist
Why Are People Affected Differently
Neuromuscular Disease; A Spectrum

Its severity varies widely within the Myotubular and Centronuclear Myopathy Patient Population
What Are The Respiratory Complications in MTM?
Respiratory Complications in NMD

General muscle weakness

- Decreased chest wall compliance
  - Shortening of the chest wall and respiratory muscles
    - Decreased tidal volumes
  - Hypoventilation

- Ineffective cough
  - Recurrent chest infections
Respiratory Complications in NMD

General muscle weakness

- Decreased chest wall compliance
  - Shortening of the chest wall and respiratory muscles
  - Decreased tidal volumes

Respiratory muscle weakness

- Hypoventilation
  - Optimal Ventilatory Support

Ineffective cough

- Recurrent chest infections
Effects of NIV: Hospital admissions


59 children with nocturnal hypoventilation

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<td>Hospital (days/year)</td>
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<tr>
<td>Pre</td>
<td>41.7 (0 to 82)*</td>
<td>48 (10.5)#</td>
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<td>Post</td>
<td>2.3</td>
<td>8.1 (2.9)#</td>
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*Range, #SEM, mean±SD+
How Can I Maintain My Lung Function?

- General muscle weakness
  - Decreased chest wall compliance
    - Shortening of the chest wall and respiratory muscles
      - Decreased tidal volumes
  - Respiratory muscle weakness
    - Hypoventilation
      - Ineffective cough
        - Recurrent chest infections
      - Maintaining lung function
Effects of NIV: Chest Wall Stretching

Effects of NIV: Chest wall stretching

(Chatwin et al., Arch Dis Child, 2011)
Lung Volume Recruitment

Lung volume recruitment bag

Alpha 301

Bird Mark 7

Intersurgical

Salva

Care Fusion
Air Stacking

Lung volumes

Hold!
1-2-3

Breath in

Air exhaled

MIC
(maximal insufflation capacity)
/ LIC
(lung insufflation capacity)

Thanks to Tiina Andersen
Respiratory Complications in NMD

General muscle weakness

- Decreased chest wall compliance
  - Shortening of the chest wall and respiratory muscles
  - Decreased tidal volumes

- Respiratory muscle weakness
  - Hypoventilation

- Ineffective cough
  - Recurrent chest infections

Management of chest infections
Respiratory Complications in NMD

- Medical Management
- Management of Chest Infections
  - Secretion mobilizing techniques
  - Cough augmentation techniques
What Is The Correct Order For My Inhaled Medication?

(Rochester and Chatwin, ERS buyers guide, 2012)
Secretion Movement

(Fink, Respir Care, 2007)

Normal ciliary beat frequency: 10-15Hz (Chilvers et al., Thorax 2003)

• Viscosity
• Depth of liquid layer
• Elasticity
• Airflow equilibrium
• Position of airway / gravity
Hypertonic Saline vs Normal Saline: What’s The Difference? & What’s The Max Strength You Would Use?

- Increases amount of sodium (salt) in the airways. Salt attracts water into the airways, which thins the mucus, making it easier to clear.
## Hypertonic Saline vs Normal Saline: What’s The Difference? & What’s The Max Strength You Would Use?

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<tr>
<th></th>
<th>0.9%</th>
<th>3 or 3.5%</th>
<th>7%</th>
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<td>Normal Saline</td>
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<td>Hypertonic Saline</td>
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<tr>
<td>Hypertonic Saline</td>
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Does Salbutamol Help?  
Can You Give This To Young Infants?  
In What Situation (Unwell or Routinely)?

• Why are we giving it?  
  – Airway irritability / Bronchoconstriction?  
    • Yes  

• Routinely?
How To Manage Bacteria Colonisation?

- **1st step** is irradiation with oral antibiotics
- **2nd step**? prophylactic oral antibiotics
- Or Nebulised antibiotics
- **Best deposition**
Respiratory Physiotherapy?
Where Are The Secretions?

- Secretion mobilising
- Cough augmentation
Respiratory Physiotherapy

Airway Clearance Techniques (ACT’s)

Proximal ACT’s "Cough Augmentation"
- Assisted Inspiration
  - Single Breaths
    - Mechanical Insufflation
    - Non Invasive Ventilation (NIV)
    - Intermittent Positive Pressure Breathing (IPPB)
  - Stacked Breaths
    - Air Stacking (AS)
    - Glossopharyngeal Breathing (GPB)
    - Volume Cycled NIV
    - Lung Volume Recruitment Bag
    - Resuscitation Bag with Patient Holding Their Breath
- Manual Assisted Cough (MAC)
  - Mechanical Exsufflation

Assisted Expiration
- Any Combination of Assisted Inspiration and Assisted Expiration Techniques
- Mechanical Insufflation Exsufflation (MI-E)

Assisted Inspiration and Expiration

Peripheral ACT’s "Sputum Mobilizing"
- Manual Techniques (MT)
  - High Frequency Chest Wall Oscillation (HFCWO)
  - High Frequency Chest Wall Compression (HFCWC)
  - Intrapulmonary Percussive Ventilation (IPV)
  - Chest Wall Strapping (CWS)

(Toussaint et al, Neuromuscular Disorders, 2018 & Chatwin et al, Respiratory Medicine, 2018)
Peripheral Airway Clearance Techniques; Chest Clapping, Vibration, and Shaking

(Wong et al., JAP, 2003)

Increases expiratory tidal volume during vibrations
How Long Should You Give Chest Percussion For & Why?

- No more than 30 seconds and then rest for 20-30. Repeat as required but usually for a minimum of 5 minutes each area
- Doing deep breaths or adequate ventilation and percussion for no more than 30 seconds does not lead to desaturation (Pryor JA et al. Thorax, 1990)
What’s The Best Technique For Vibrations in Adults?
What If You Have Small Hands!
Do Manual Techniques Work?
B

Manual techniques/positioning

(Toussaint et al, Neuromuscular Disorders 2017)
When To Use Peripheral ACT Like The Vest™
HFCWO

- High secretion demand within the lungs and not central airway
- Long treatment times
- Recurrent chest infections despite optimal treatment
A

High frequency oscillations/percussions

(Toussaint et al, Neuromuscular Disorders 2017)
MI-E Devices with Oscillations
(Sancho, Respir Care, 2016)

No Oscillations

Oscillations on Insufflation

Oscillations on Exsufflation

Oscillations on Insufflation & Exsufflation
Modified Autogenic Drainage / Chest Wall Strapping

(Schoni, J R Soc Med, 1989)

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<th>Collecting</th>
<th>Evacuating</th>
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<td></td>
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<tr>
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<td>FRC</td>
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- Low lung volume
- Mid lung volume
- High lung volume

![Image of a patient receiving chest wall strapping](image-url)
Respiratory Physiotherapy

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"Cough Augmentation"

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- Assisted Inspiration and Expiration
  - Any Combination of Assisted Inspiration and Assisted Expiration Techniques
  - Mechanical Insufflation Exsufflation (MI-E)

Peripheral ACT's
"Sputum Mobilizing"

- Manual Techniques (MT)
  - High Frequency Chest Wall Oscillation (HFCWO)
  - High Frequency Chest Wall Compression (HFCWC)
  - Intrapulmonary Percussive Ventilation (IPV)
  - Chest Wall Strapping (CWS)

(Toussaint et al, Neuromuscular Disorders, 2018 & Chatwin et al, Respiratory Medicine, 2018)
Assessment of Cough

Inspiration

- Inspiratory muscle weakness

Glottic function

- Artificial airway
- Bulbar dysfunction

Expiration

- Expiratory muscle weakness
e.g. SCI, SMA
Peak Cough Flow (PCF)  
(McCool, Chest, 2006)

1. Full face mask
2. Pediatric flow meter (more accurate at low flows)
What Technique & When?
Recommendations For Proximal ACT’s Dependent on PCF, FVC or FVC%Pred

(Toussaint et al., Neuromusc Disord, 2017)
Cough Augmentation

PCF > 360 L/min
- No assistance
  - Lung volume recruitment bag
  - Intersurgical
  - Alpha 301
  - Bird Mark 7

PCF < 160 L/min
- Breath stacking
  - IPPB
  - NIV
  - MAC
  - MI-E
  - +/- MAC

- Intersurgical
- Cough Assist E70
- Care Fusion
- Philips Respironics
- Pegaso
- B&D Electromedical
- NiPPY Clearway
- Dima italia
Mechanical Insufflation-Exsufflation (MI-E)
When To Use Mechanical Insufflation-Exsufflation (MI-E) (Cough Machines)

• Questions
  – It depends if you need one?
  – What is your PCF?
  – Have you tried any other techniques?

• Answers
  – Recurrent chest infections despite other techniques – yes
  – Lots of secretions – yes
  – Weak – yes
  – Tracheostomy - possibly
MI-E- How Many Cycles/Coughs Would You Recommend?

- Daily or twice daily airways clearance for all
  - Why to be familiar with the device
  - Evaluate effectiveness
  - Check chest
  - Maintain a compliant chest wall (stretch)

- Cycles and coughs?
  - Depends on how your device is set up
Can Infants Have MI-E More Frequently When Unwell Or Should You Stick To Twice A Day?

- Use when SaO2 < 95%
- Use as much as needed!
- Beware of fatigue and rest on ventilator
How Young Can You Start MI-E In A Infant?

• The youngest is 3 months
  – Sensible settings
• From 12 months onwards seems to get best co-ordination
Is There A Maximum Number Of MI-E Cycles You Can Give To A Infant?

• No
  – You need to give rests in-between and have suction ready as they wont clear into mouth

• I might do up to ten and then suction
  – Depends when you feel ratterling in the tubing
Aerophagia With MI-E

- Options
  - Vent through a gastrostomy
  - Try it in a more upright position
  - Rest longer in-between
  - Try to let the machine take over
  - Decrease insufflation pressure
  - Check timings possibly increase pause
  - Little and often if able
Other considerations?
Equipment Required In The Management of MTM

<table>
<thead>
<tr>
<th>Mask choice start with nasal</th>
<th>2 x Appropriate ventilator for the child and interface</th>
<th>Ancillary equipment</th>
<th>Education and training</th>
</tr>
</thead>
</table>

- Custom made

- With battery
Workshop report

228th ENMC International Workshop:
Airway clearance techniques in neuromuscular disorders
Naarden, The Netherlands, 3–5 March, 2017
Michel Toussaint a,*, Michelle Chatwin b, Jesus Gonzales c, David J. Berlowitz d the ENMC Respiratory Therapy Consortium

Respiratory Medicine 136 (2018) 98–110

Contents lists available at ScienceDirect

Respiratory Medicine

journal homepage: www.elsevier.com/locate/rmed

Review article

Airway clearance techniques in neuromuscular disorders: A state of the art review

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