LUNG VOLUME RECRUITMENT IN NEUROMUSCULAR DISEASE

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Objectives

• To distinguish lung volume recruitment techniques and explain their application in neuromuscular disease.
• To appraise the evidence in the literature for benefit of lung volume recruitment in individuals with neuromuscular disease.
• To compare the role of lung volume recruitment therapy in clinical care guidelines for adults and children with neuromuscular disease.
The Clinical Problem

- Respiratory complications are primary cause of morbidity and mortality
  - Hypoventilation
  - Recurrent pneumonia/atelectasis

- No definitive treatment for underlying disease

- **Management goals:**
  - Slow decline in lung function
  - Avoid life-threatening respiratory infections
  - Improve quality of life
Case

- 14 y.o. with DMD
- Seen in clinic after a 3 year absence
- Now full-time in wheelchair
- 2 pneumonias last year requiring antibiotics, hospitalized for one
- Daytime fatigue, morning headaches, less energy
- “Gurgling in throat”, “difficulty coughing stuff up”
- FVC ↓ 70% to 40%, weak cough
How LVR Works

- Device that allows breath-stacking by applying slow inspiratory pressure causing lung inflation

Bedard & McKim, Ontario Thoracic Reviews, 2012
<table>
<thead>
<tr>
<th>Devices for LVR</th>
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<tbody>
<tr>
<td><strong>Self-inflating AMBU-bag with one-way valve</strong></td>
<td><strong>Respironics In-exsufflulator</strong> (Cough Assist)</td>
</tr>
<tr>
<td>Direct feedback given to provider → comfortable volume delivered</td>
<td>Mechanically provides positive pressure breaths followed by rapid negative pressure which generates a cough</td>
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<tr>
<td>May need abdominal thrust to generate cough</td>
<td></td>
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<tr>
<td>Readily available</td>
<td>Not covered by all provincial insurance plans (support in Ont., Que., BC)</td>
</tr>
<tr>
<td>Inexpensive ($70)</td>
<td>Expensive (~ $4500-6000)</td>
</tr>
<tr>
<td>Portable</td>
<td>Cumbersome, less portable</td>
</tr>
<tr>
<td></td>
<td>Better studied</td>
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Procedure

• Via facemask or mouthpiece positive pressure applied using an AMBUbag with one-way valve or CoughAssist device

• Pressure settings determined by visual inspection of chest wall and patient comfort

• 3-5 breaths, coordinated with patient’s respiratory pattern, may be followed by spontaneous or manually assisted cough (abdominal thrust) while at MIC

• Total of 3-5 cycles, as tolerated

• Followed by suctioning PRN
Theoretical Benefits

• Improves cough efficacy
  • ↑ inspired volume and expiratory forces applied to the inspired volume; both ↑ expiratory flow rates during coughing
  • Measured as Peak Cough Flow

• Maintains chest wall compliance and range of motion
  • Measured as Maximum Insufflation Capacity (MIC)

• Maintains lung compliance
  • ? Slows progression of restrictive lung impairment (FVC)

• Aids airway clearance
  • Even if secretions get into larger airways without an effective cough they cannot be expelled
However…

No RCTs, nor any long-term controlled prospective studies evaluating LVR as a treatment in neuromuscular disease
Risks/Complications - Adults

- Two adult patients (one with DMD) who had pneumothorax while using mechanical insufflation
  - Other risk factors present: NIV, mild COPD
    - Suri, AJRCCM, 2008

- One case of pneumothorax with LVR
  - 73 year old woman with asthma and 20 pack year smoking
    - Westerman, J Bras Pneumol., 2013
Risks/Complications - Children

- No reports of pneumothorax directly attributable to insufflation-exsufflation devices in children
  - Hull, Thorax, 2012

- One case of intestinal perforation with LVR
  - 9 month old with trisomy 8
  - annular pancreas (non-compliant second part of duodenum?)
    - Dwight, J Ped Surg, 2004
Evidence
Evidence: ↑Cough Efficacy

- Most studies used the Emerson In-exsufflator, in combination with other treatments
  - Difficult to assess the impact of LVR alone

- Insufflation with a bag and mask ↑ peak cough flow as compared to unassisted cough, to a level comparable to mechanical insufflation

- PCF achievable with LVR is 1.8 times greater than with unassisted cough
  - Bach, Chest, 1993

- With mechanical in-exsufflation, PCF in a clinically effective range can be achieved
  - Chatwin, ERJ, 2003; Sancho, Chest, 2004
Evidence: ↓ Restriction

- LVR aims to maintain “range of motion” of chest wall by preventing atelectasis and chest wall contractures
- One case series
  - 0.5-24 years of follow-up
  - N=282 with neuromuscular disease (mostly adults)
- Maximum insufflation capacity (MIC) ↑d despite ↓ vital capacity
  - Max volume of air that can be held in the lungs with a closed glottis after breath-stacking

Evidence: ↓ Restriction

• Retrospective study in Multiple Sclerosis
• n=79, 35 with > 1 visit, prescribed LVR BID if FVC < 80% pred

• LVR was associated with slower rate of decline of FVC and PCF over median 13 months

• FVC & PCF rate of decline were slower if PCF improved at first visit

• Success of LVR was not associated with age, sex, disease-modifying treatment or meds for spasticity/kyphoscoliosis

  • Srour, PLOS One, 2013
Evidence: ↓ Respiratory Infections

• Integrating in-exsufflator into plan of care has been successful at:
  - ↓ hospitalization
  - ↓ pneumonia
  - ↓ episodes of respiratory failure
  - ↓ tracheostomy


BUT – difficult to determine whether benefits are due to NPPV, in-exsufflator, or both
Long-term Studies
Long-term LVR Use - Adults

- Retrospective cohort study of PFT trajectory in adults with DMD pre & post LVR

FVC Rate of decline:
Pre-LVR = -4.7% predicted/yr
Post LVR = -0.5% predicted/yr
(p < 0.001)

McKim & Katz, Arch Phys Med Rehab, 2013
Long-term LVR Use - Kids

- Only pediatric study is retrospective
- Regular use: daily – q4hours
- N=62, neuromuscular disease and impaired cough, age 3 months-28.6 years
- Median duration of use = 13.4 months
- 6% had improved chronic atelectasis
- 8% had ↓ frequency of pneumonias
- Total # of respiratory infections was too low to permit meaningful comparison with pre-treatment period

Miske, Chest, 2004
Registry Data

- Analysis of patient registry of SMA Type 1
- Comparison of patients born 1995-2006 vs. 1980-1984
- 70% reduction in risk of death
- Controlling for demographic and clinical care variables
  - Use of a mechanical in-exsufflator had a significant independent effect in reducing death
    - Oskoui, Neurology, 2007
Current Guidelines
Broad Statements

- Patients should be taught airway clearance techniques and how to employ them early and aggressively
  - ATS, 2004

- Children with weak cough should be taught airway clearance
  - British Thoracic Society, 2012

- Techniques require substantial acclimatization - should be trained when the patient is well, prior to an acute infective requirement
  - Australian guidelines, 2010
Specific Thresholds

• DMD Care Considerations Working Group (CDC)
  
  • Manual or mechanical LVR when FVC < 40% predicted
  
  • Mechanical in-exsufflation when:
    • Respiratory infection & PCF < 270 L/min
    • Baseline PCF < 160 L/min or MEP < 40 cm H$_2$O
    • Baseline FVC < 40 % predicted or < 1.25 L in older teenagers/adults
  
  • Bushby, Lancet Neurol, 2010
Specific Thresholds

- **Australian Guidelines**
- Baseline assisted PCF < 270 L/min - likely to ↓ to < 160 L/min during chest infections so should have access to insufflation/mechanical in-exsufflation
  - Training of insufflation should commence when VC < 2L or 50% predicted
  - For patients with VC < 1 to 1.5L, insufflations should precede manual assisted coughing techniques (e.g. abdominal thrusts).
    - German Society for Pneumology, 2010

- **German Guidelines**
- PCF < 270 L/min → start air stacking, then manual assisted cough, then insufflator/exsufflator
  - German Society for Pneumology, 2010
Device Recommendations

- Manual cough assist and air-stacking methods to achieve MIC are effective to improve cough efficiency and should be used when appropriate.

- Mechanical in-exsufflation should be used if very weak, loss of bulbar function or not cooperative with manual technique
  - British Thoracic Society, 2010

- Manual or mechanical LVR
  - DMD Care Considerations, 2010

- Step-based secretion management: air stacking/manual hyperinflation → assisted cough techniques → mechanical cough assistants
  - German Society for Pneumology, 2010
Pressure Settings

• For Mechanical Inexsufflation

• **Adults:** +/- 40 cmH$_2$O appear to safely provide adequate PCF for the majority with neuromuscular disease.
  • Australian Guidelines, 2010

• **Children:** usual range +/-15-40 cmH$_2$O, although some recommend +/-35-50 cmH$_2$O
Caveats

- Mechanical in-exsufflation can be ineffective in patients with very poor bulbar dysfunction with insufflation capacity >1L, where dynamic airway collapse occurs.
  - ACI Respiratory Network, 2010
CTS Pediatric Home Mechanical Ventilation Guidelines (in development)

- Airway clearance techniques should be taught to children and caregivers as a preventative strategy in those with impaired cough

- Manual LVR for patients with impaired cough
  - clinical assessment
  - MEP < 60 cm H$_2$O
  - CPF < 270 L/min
  - FVC <40 % predicted

- Manual and/or mechanical LVR recommended during respiratory infections
Current Standards of Care

• Given lack of studies, implementation limited

• Several groups worldwide have called for further study before widespread adoption

• Listed as top research priority in ATS and recent British Thoracic Society Guidelines
  • Finder, AJRCCM, 2004; Hull, Thorax, 2012
Canadian Survey of Pediatric Respiratory Management in DMD (2010)

- Response rate 66%, including all provinces where Ped Resp practice
- 18% (N=6) of Respirologists (at 3 centres) use LVR BID during clinical stability, in 33% of their patients
  - Katz, Pediatr Pulmonol, 2012
Clinical equipoise exists. Given additional care burden, this warrants further study...
STEADFAST Study

• **Stacking Exercises Attenuate the Decline in FVC and Sick Time (STEADFAST)**

  • 14 centre RCT across Canada
  • Boys 6-16yrs with DMD and FVC > 30% predicted
  • Randomized to usual care or usual care + LVR BID with self-inflating resuscitation bag and one-way valve
  • **Primary outcome**: rate of FVC decline (% pred) over 2 years
  • **Secondary outcomes**: hospitalizations, ICU days, quality of life, adherence to LVR therapy, PCF, MIC, MIP/MEP
Case

• **LVR** technique taught – to be used twice daily
• Sleep study showed severe hypoventilation – started on **nocturnal NPPV**

**Over the subsequent year:**
• Able to clear secretions – no more gurgling!
• Handled 3 colds well – no pneumonias!
• Increased daytime energy, no more headaches!
• “feels much better”
Take-Home Messages

• LVR is considered standard of care in some institutions, but widespread use is limited by lack of familiarity with device and evidence-based clinical evaluation

• LVR may slow the decline in lung function in children with NM disease

• Further studies may demonstrate the benefits of this therapy, which would result in more widespread, standardized use of this tool for treatment of NM disease