Respiratory Management in Motor Neurone disease

Dr Denise Gibson
Consultant Physiotherapist
University Hospital Southampton NHS Foundation Trust
Respiratory Failure

Inability to Ventilate
Inspiratory Muscle Weakness

Aspiration Risk
Upper Airway Glottic Muscle Weakness

Neuromuscular Respiratory Failure

Inability to cough
Inspiratory/Expiratory and glottic muscle weakness
Evolution of ventilatory decompensation in Neuromuscular disease

- Normal breathing
- REM related sleep disordered breathing
- Daytime ventilatory failure
- NREM and REM sleep disordered breathing

Factors:
- FVC < 20%
- FVC < 40%
- Chest infection
- NIV

Questions:
- ?
Indications for Ventilation in MND

- Prevention of respiratory decompensation
- Chest infection
- Rest Resp muscles
- Control nocturnal hypoventilation with or without symptoms
- Treat established hypercapnic ventilatory failure
- Palliation of symptoms
Symptoms of Respiratory Failure in MND

• Dyspnoea
• Excessive daytime sleepiness
• Lethargy
• AM headaches
• Orthopnoea
• Sleep disordered breathing
Assessment for NIV

- ABGs PH<7.35 (PH is main indicator for survival)
- BTS guidelines
- Baseline ABGs
- Spirometry
- Oximetry (overnight)
- Symptoms of respiratory failure
Functional evaluation of MND Probable need for NIV

- VC
- Falls by >20 to 40% from sitting to supine
- <1.2 to 1.5L
- <40% predicted
- Nocturnal hypoventilation
- Expiratory Muscle weakness
Consensus Conference: Clinical Indications for NIV in CRF

Neuromusculoskeletal disease

• Diagnostic certainty
• Symptoms eg. fatigue, dyspnoea, headaches

1 of the following:
• PaCO2 > 6.0 kPa
• Nocturnal SaO2 <88% for > 5 minutes
• MIP < 60cmH2O or FVC < 50% predicted in progressive disorders
Barriers to NIV

- Bulbar dysfunction
- Interface tolerance
- Fear of dependency
Nasendoscopy during NIV
The Larynx

Right

Back of Neck

Left

ABducted or Vocal Cords Apart

TVC

ADducted or Vocal Cords Together

Front of Throat
Effects of Positive pressure in patient with Bulbar MND
Laryngeal events during positive and negative pressure

- Laryngeal Level Adverse Laryngeal Response During MI-E
- Glottic True vocal folds Adduction of true vocal folds during insufflation; paradoxical movement of true vocal folds during inhalation creating either a slim glottic opening or a total closure of glottis

- Hypopharyngeal Tongue base Backward movement of the tongue base during insufflation that constricts the laryngeal entrance

- Hypopharynx A severe hypopharyngeal narrowing during exsufflation

(Tiina M Andersen et al Respir Care 2018;63(5):538–549.)
what are the potential benefits of using NIV in the treatment of neuromuscular disease?
NIV in NMD

- Simmonds et al 2000
  - Improved life expectancy by 10-12 years in DMD
  - Eagle et al 2002
  - Improved QOL

- Nickol et al 2002
  - Increase in insp and expy and transdiaphragmatic muscle strength in NMD

- Ward et al 2003
  - Increased QOL
Effect of NIV on sleep quality

Influence of noninvasive ventilation (NIV) on sleep-breathing and sleep

<table>
<thead>
<tr>
<th></th>
<th>Before NIV</th>
<th>During NIV</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>RDI·h⁻¹</td>
<td>10.5±13.1</td>
<td>3.1±3.5</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>REM-RDI·h⁻¹</td>
<td>20.5±21.1</td>
<td>3.0±5.3</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Arousal index·h⁻¹</td>
<td>20.6±14.3</td>
<td>10.2±3.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Light-sleep %</td>
<td>55±12</td>
<td>44±13</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Slow-wave-sleep %</td>
<td>24±9</td>
<td>34±9</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>REM-sleep %</td>
<td>18±6</td>
<td>20±6</td>
<td>0.18</td>
</tr>
</tbody>
</table>

RDI: respiratory disturbance index per hour sleep; REM-RDI: respiratory disturbance index per hour rapid eye movement (REM)-sleep; arousal index: electrocephałohographic arousals per hour sleep.
Survival: Probability of continuing domiciliary NIV
NIV in MND/ALS: Quality of life

- Bourke et al AJCCRM 2001: Assessment pre, 1, 3, 5 month after starting NIV

- Generic: Improvements in GWbS ($p=0.039$), SF36 emotional limitation, health perception

- Specific: Improvements Epworth SS, CRDQ dyspnoea, fatigue & mastery

- Improvements at 1 month maintained at 5 months despite disease progression

- Indices of sleep-related symptoms most responsive
NIV in MND/ALS: Quality of life

- *Lyall et al Neurol 2001*: NIV increased Vitality domain (SF36) by 25% for up to 15 months despite disease progression
NIV: Quality of life

• Effect of non-invasive ventilation on survival and QOL in patients with amyotrophic lateral sclerosis: a randomised control trial

• Bourke SC, John Gibson et al
• Lancet Neurology 2006;5:140-147.
92 patients assessed every 2 months

- Randomised when
  a) Orthopnea
  or
  b) MIP < 60% predicted
  or
  c) Symptomatic hypercapnia

NIV n=22, standard care n=19
Outcomes

- Group 1 Better bulbar function (n=20)
  NIV Improved survival (mean 205 days, p<0.01)
  Improved QOL

  Group 2 Poorer bulbar function (n=21)
  No survival benefit, inconsistent effect on QOL.
Survival and lung function in MND/ALS with NIV
Kleopa et al 1999 J Neurol Sci

NIV use  Group 1 > 4hrs  Group 2 < 4 hrs Group 3 Nil
Survival  Gp 1: 14.2 mths  Gp 2: 7.0 mths  Gp 3: 3.0 mths
Mechanisms of action of NIV

• Effect on sleep disordered breathing?
• Effect on respiratory muscle strength?
• Effect on chest wall & lung mechanics?
• Effect on ventilatory drive/chemosensitivity?
• Combination of above
Inspiratory, expiratory and transdiaphragmatic muscle strength (restrictives)

Nickol et al. Thorax 2005

- Inspiratory RMS (cmH\(_2\)O)
- Expiratory RMS (cmH\(_2\)O)
- PiMax
- Poes
- SnIP
- Pgas
- PeMax

Day 0, Day 5, 3 Months

Pdi (cmH\(_2\)O)

Sniff Pdi
p = 0.76

TwPdi
p = 0.36
Efficacy of NIV: how much?
Nickol et al 2005

- NMD
- X < 4hrs NIV
NIV: Daytime Vs Nocturnal use?

- Schonhofer et al 1997
- Daytime mechanical ventilation was equally effective in improving sleep hypoventilation and architecture as nocturnal ventilatory support
- Study carried out in hospital setting with supervision.
- Larger randomised controlled trials needed
NIV: Which Interface?

- Nasal Masks
- Full face masks
- Nasal Pillows
- Mouth piece
- Tracheostomy
Interfaces: Nasal
Interfaces: Full face mask
Interfaces: Nasal Pressure sores

- Prevention
- Detax Moulding
- Towelling Tape
- Use alternate interface
Interfaces: Full Face Mask
Interfaces: Oral
Interfaces - Acute care
Tracheostomy

Ethical issues
Informed decision
Advantages for sputum management
No nasal sores
Patient can maintain speech
UHS Home ventilation service

- 400 patients
- Including tracheostomy & mask ventilated patients

Regional service across Hampshire/Dorset/Wiltshire
University Hospital Southampton

- 24 hour technical / ventilator support
- Home managed patients
- In/outpatient Ventilation trials
- Home sleep assessments
UHS Home ventilation Service

Prevalence of use of home NIV by year and indication

- Thoracic
- NM
- OHS
- COPD
- CF
- ILD/Other

Year:
- 2004
- 2005
- 2006
- 2007
- 2008
- 2009
- 2010
- 2011
- 2012
- 2013
- 2014
- 2015

Values:
- 0
- 50
- 100
- 150
- 200
- 250
- 300
Data Downloads

Pressure (cmH2O)

Patient Triggered Breaths (%)

Sleep Therapy Flags

Leak (LPM)
The future

- Remote Monitoring in Home ventilation to include Patients with MND
- Attend anywhere video clinics
Cough and Secretion clearance

Mucociliary escalator
Cough

Cough phases

- Deep Inspiration
- Glottic Closure and reopening
- Contraction of expiratory muscles
Measuring cough

- Peak Cough Flow

160l/min threshold for Mechanical assisted cough
Cough Augmentation
Simonds 2009

Figure 8.6 Schematic representation of the management for cough augmentation.
Cough

• Insufflation
• Breath stacking
• LVR Bag
Cough

• Exsufflation:
  – Manual assisted cough
Cough

Mechanical assisted cough
Thank you for Listening