Motor Neurone Disease –NICE to manage
Management of ineffective cough

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Content

- NICE guideline recommendations
- Respiratory involvement in MND
- Physiology of cough
- Objective measurements of respiratory muscle strength
- Treatment options for cough augmentation
- References
1.13 Cough effectiveness

• 1.13.1 Offer cough augmentation techniques such as manual assisted cough to people with MND who cannot cough effectively. [new 2016]

• 1.13.2 Consider unassisted breath stacking and/or manual assisted cough as the first-line treatment for people with MND who have an ineffective cough. [new 2016]

• 1.13.3 For people with bulbar dysfunction, or whose cough is ineffective with unassisted breath stacking, consider assisted breath stacking (for example, using a lung volume recruitment bag). [new 2016]

• 1.13.4 Consider a mechanical cough assist device if assisted breath stacking is not effective, and/or during a respiratory tract infection. [new 2016]
## Respiratory involvement in NMD

<table>
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<tr>
<th>Muscle condition</th>
<th>Frequency of respiratory involvement in hereditary neuromuscular conditions</th>
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<td>Duchenne muscular dystrophy</td>
<td>Inevitable</td>
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<td>Spinal muscular atrophy type 1</td>
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<td>Spinal muscular atrophy type 1 with respiratory distress (SMARD 1)</td>
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<td>Motor neurone disease</td>
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<td>Acid maltese deficiency</td>
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<td>Limb girdle muscular dystrophy type 2C, 2D, 2F, 2I</td>
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<td>Myotubular myopathy</td>
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<td>Congential muscular dystrophy</td>
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<td>Limb girdle muscular dystrophy type 1, 2A, B, G, H</td>
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<td>Emery dreifuss muscular dystrophy</td>
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<td>Mitochondrial myopathy</td>
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<td>Central core disease</td>
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<td>Limb girdle muscular dystrophy type 1,2A,B, G, H</td>
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<td>Occassional</td>
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Respiratory complications in MND

- Respiratory muscle weakness
  - Decreased chest wall compliance
  - Ineffective cough
  - REM related nocturnal hypoventilation
- Shortening of respiratory muscles and chest wall deformity
- Recurrent chest infections
- NREM and REM related hypoventilation
- Decreased tidal volumes
- Daytime hypercapnic ventilatory failure
Physiology of Cough

3 phases of cough combine to produce adequate PCF (> 360L/min)

Inspiratory

- Active abduction of the glottis supports inspiratory airflow
- Rapid inhalation due to diaphragm and inspiratory accessory muscle contraction
- Lengthens the expiratory muscles creating greater force

Compressive

- Simultaneous adduction of glottis and expiratory muscle contraction
- Compresses alveolar gas volume

Expiratory

- Glottis opens
- Release of compression; central airway pressure drops
- Pleural and alveolar pressure rise
- High linear velocity in airways creates a shearing effect on the mucus

Boitano 2006
Impact of Neuromuscular Weakness on Cough

**Inspiratory**
- In pts with bulbar weakness glottis function impaired
- Weak diaphragm and inspiratory muscles mean weak inspiration
- Poor inspiratory volume produces minimal stretch of exp muscles, decreasing length tension

**Compressive**
- Poor glottis closure limits ability to build up compressive forces, huffing may be required.
- Expiratory muscle contraction weak
- Poor compression of alveoli

**Expiratory**
- Glottis opens
- Release of compression; central airway pressure drops, but due to decreased exp strength reduced flow rate
- Pleural and alveolar pressure rise
- Lower linear velocity of air in airways creates a shearing effect on the mucus but less effective

Boitano 2006
Ineffective cough

- PCF <160 L/min or >160 L/min but < 270L/min

- Inspiratory phase:
  - weakened respiratory muscle as course of the disease
  - Furthered weakened by acute respiratory infections
  - Unable to fill their lungs with enough air

- Compressive/Glottis Closure:
  - Bulbar involvement reduces this ability
  - Unable to generate the pressure required

- Expiratory Phase:
  - Unable to generate effective expiratory flow
Respiratory Management

- Assess VC/SNIP/PCF
- Assess of cough
- Optimise respiratory care in event of retained secretions/bronchitis infection
  - Humidification
  - Bronchodilators
  - Mucolytics
  - Physiotherapy techniques
- **Caution with O2**
- Consider medical management including
  - Saliva management
  - Low threshold for antibiotics
Optimal medical management

• Flu and pneumonia vaccination
• SALT and nutritional assessment
• Optimal management of oral secretions
• Optimal reflux management if reflux is suspected or an issue
• Low threshold for Abx
• Early detection of chest infections
Forced Vital Capacity

- Normal Values 3-6 litres
- FVC > 1.5 litre required for effective cough
- A change in FVC by 25% lying down indicates diaphragm weakness

Miller et al 1999

Good Practice Points

- > monitor vital capacity in patients with neuromuscular disease to guide therapeutic intervention
- When VC falls to <50% take appropriate action to minimise the risk of respiratory failure and cough impairment

Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient, 2009

However this marker is non-linear and less predictive of sleep disordered breathing in MND that direct measurement of inspiratory muscle strength.

Lyall et al 2001
Peak Cough Flow (PCF)

- Normal values >350l/min to 600l/min
- PCF >160l/min required to clear secretions
- PCF >270l/min when well has little risk of respiratory failure during RTI
Cough and airway secretion management: Peak Cough Flow

Recommendation:

- Peak cough flow should be measured regularly in patients with neuromuscular disease
- PCF > 160l/min sufficient
- PCF < 160l/min ineffective
- PCF > 270l/min (assisted or unassisted) when well have little risk of developing respiratory failure during a respiratory tract infection
- PCF must be greater than the critical level of 160l/min when the patient is clinically stable to avoid the risk of respiratory failure during an infection.

- PCF < 270l/min in a medically stable patient, introduce strategies for assisted airway clearance to raise it above 270l/min
- PCF < 160: additional strategies to assist secretion clearance must be used
- If PCF remains equal to or less than < 160l/min despite additional strategies, contact medical colleagues to discuss ventilation and/or airway management needs.
Sniff Nasal Inspiratory Pressure (SNIP)

- Normal values: males >-70cmH2O, females >-60cmH2O
- 32% of predicted value has a 81% sensitivity for hypercapnia in ALS. Lyall et al 2001
Neuromuscular patients admitted to KCH.

Ineffective Cough (PCF < 150 l/min)

PCF ≥ 170 l/min but > 160 l/min

PCF > 160 l/min = effective cough
No intervention required.

Does the patient have secretions?

No
Yes

Does the patient tire with Manual Assisted Cough?

No
Yes

Is the patient unable to increase PCF with Manual Assisted Cough?

No
Yes

Trial Lung Volume Recruitment bag and consider for home use.

However...
The Cough Assist MUST be considered in cases where there is no improvement in PCF above 150 l/min.
Assessment of cough should be part of the routine, and treatment targeted to findings.

Increasing Muscle weakness

Combination of inspiratory and expiratory techniques

Mechanical In-ex sufflator (cough assist)

Combination of in-ex sufflator and insp/exp techniques

Increasing Muscle weakness

Worsening of PCF

Inspiratory techniques
- Breath stacking
- NIV
- IPPB

Expiratory techniques
- Manual assisted cough (MAC)
Breath stacking

• Unaided deep breathing exercises
• Lung recruitment bag
• Glosopharyngeal breathing
• Non invasive ventilation
LVR (Inspiratory phase)

- Lung volume recruitment is used to improve peak cough flow (PCF) by reaching maximum inspiratory capacity (MIC) in patients with paralytic /restrictive disorders.
IPPB – Intermittent Positive Pressure Breathing

- The delivery of positive pressure during inspiration to a pre-set inspiratory pressure. It is patient triggered via a mask or mouthpiece. Airway pressure returns to atmospheric pressure on expiration.
Manual assisted cough
Insufflator – exsufflator / cough assist

- A mechanical insufflator – exsufflator uses positive pressure to deliver a maximal lung inhalation, followed by an abrupt switch to negative pressure to the upper airway. The rapid change from positive to negative pressure is aimed at stimulating the airflow changes that occur during cough, thereby assisting sputum clearance (Chatwin, 2009)
Practical session

Does anyone want to practice?

Any questions?
Thanks for listening.
References

• Boitano LJ. Management of airway clearance in neuromuscular disease. Respir Care. 2006 Aug; 51(8):913-22
• Kings College Hospital physiotherapy department guidelines
• Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient, Thorax, 2009 (64), BTS/ACPRC
• Miller et al. Practice parameter: The care of the patient with amyotrophic lateral sclerosis (an evidence-based review). Neurology 1999, April 52(7) 1311
• NICE guidelines NG42, Feb 2016
• Website: http://www.geronguide.com/gallery/index.php/Bronchitis/bronchitis-19
http://www.rcjournal.com/cpgs/ - AARC american association for resp care don’t recommend chest physio for NMD patients!
Guidelines for the physiotherapy management of the adult, medical, spontaneously breathing patient
Thorax 2009: 64  Joint BTS/ACRRC guideline