King's College London
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Guy's and St Thomas' NHS
NHS Foundation Trust

Lane Fox Respiratory Unit,
St Thomas' Hospital

Secretion management in Motor Neurone Disease (Bulbar/Chest)

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5th March 2024, Tunbridge Wells
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London Marathon: MND competitor to take on marathon with ventilator

www.bbc.co.uk/news/April 21st 2023

Mr Perkins will be pushed in his wheelchair by Chrissie Wellington

Overview

Sialorrhea

-Non-pharmacological

-Pharmacological management

-NICE guidelines

Chest secretion management

-Pharmacological

-Cough function assessment

-Cough assistance techniques and devices

Role of tracheostomy

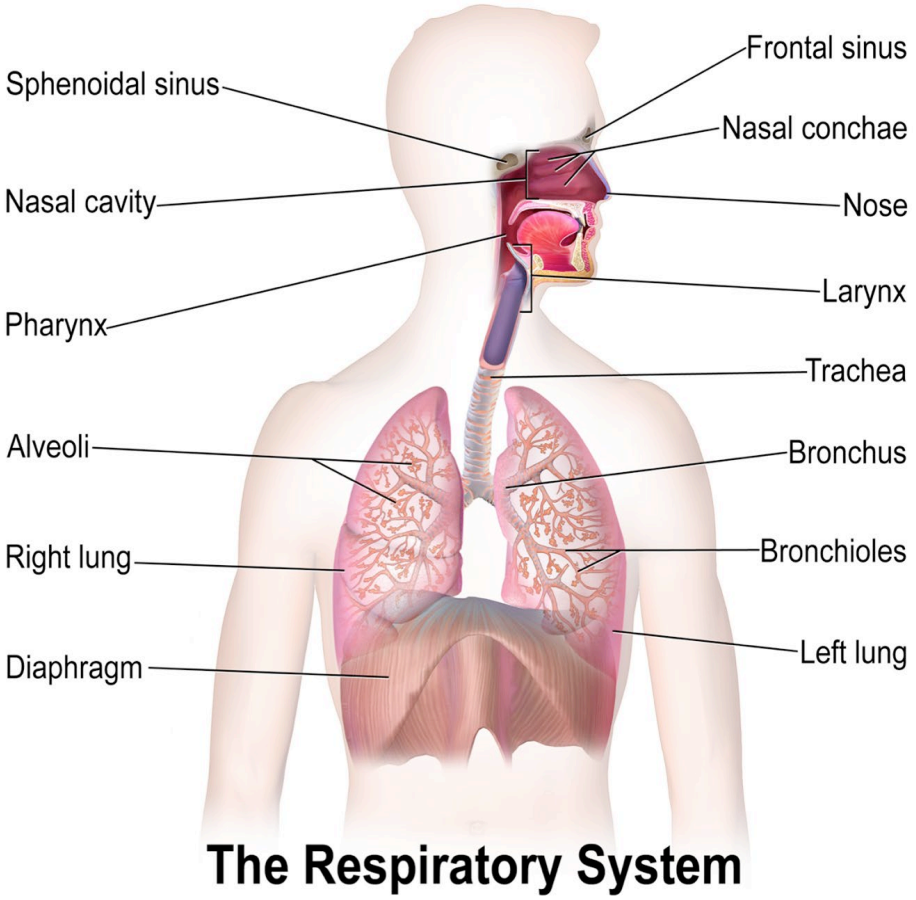
Anatomy review

Upper respiratory tract:

- Nasal Cavity
- Pharynx
- Larynx

Lower Respiratory tract:

- Trachea
- Bronchus
- Bronchioles
- Alveoli



Sialorrhea (drooling): a pratical approach

- Normal salivary production:
  - 0.5 L to 1.5 L daily
- Decreased ability to manage normal saliva production due to weakness of the tongue, lip and swallowing muscles.
- A build up of saliva and thick tenacious mucus can disrupt sleep and increase the risk of choking.

- Balance between managing drooling and drying some of the most complex to manage

Non pharmacological management

- Position body and provide head and neck support
- prevent and reduce skin irritation
- portable suction device
- oral swabbing and syringing.

Thick tenacious saliva

- Check/increase fluid intake
- natural mucolytic agents:
  - papaya enzyme, juices and ice cubes - grape, apple, pineapple and papaya
- frequent swabbing of the mouth:
  - using plain water or a mouth wash of one teaspoon bicarbonate of soda or one teaspoon salt in a glass of water, especially after meals (avoid harsh mouthwashes)
- nebulised saline may be helpful
- assisted cough technique can help

Xerostomia or dry mouth:

- Often related to mouth breathing during sleep
- Sip of fluids regularly
- Oral lubricants:
  - Aquaspray, dry mouth products from the chemist, or citrus lollies to stimulate saliva. Or try 100 parts grape seed oil to 1 part peppermint oil
- Review medications - some such as anticholinergic medications may cause dry mouth

Pharmacological Management

- Anticholinergic medications are the first-line therapy of sialorrhea. Individual medication choices should be tailored to patient factors
  - Glycopyronium preferred in the elderly as doesn't cross the blood-brain barrier
  - Hyoscine patches (preferred to medication)- **National shortages**
- Atropine drops (eye drops and off licence)
  - Short acting 50% dose excreted in 4hrs (helpful cuff down with tracheostomy)
- Amitryptyline

Pharmacological Management

- Botulinum toxin is effective for management of sialorrhea in ALS. It can be used as second-line therapy and should be considered after feeding tube insertion because of the theoretical risk of worsening swallowing or airway integrity.
- Focal salivary gland radiation is an option for management of sialorrhea as second- or third-line therapy (usually irreversible).

Cochrane Database Syst Rev. 2022; 2022(5): CD006981. PMC
Published online 2022 May 20. doi: 10.1002/14651858.CD006981.pub3

Treatment for sialorrhea (excessive saliva) in people with motor neuron disease/amyotrophic lateral sclerosis

Monitoring Editor: Cochrane Neuromuscular Group, Eleanor James, Cathy Ellis, Ruth Brassington, Sivakumar Sathasivam, and Carolyn A Young

Paucity of RCT data.

Botulinum toxin B injected into parotid and submandibular glands may have beneficial effect on patient reported outcomes and volume of saliva at 8 weeks. No clear benefit on quality of life measures were observed.

Oral administration of 20 mg dextromethorphan hydrobromide and 10 mg quinidine sulphate twice daily (Nuedexta) probably slightly improves patient-reported sialorrhea.

Lack of RCT evidence for hyoscine patches in a cross-over design of versus Botulinum A/ Radiotherapy. Too small numbers.

NICE guidelines summary

Saliva problems

1.8.10 If a person with MND has problems with saliva, assess the volume and viscosity of the saliva and the person's respiratory function, swallowing, diet, posture and oral care. **[2016]**

1.8.11 If a person with MND has problems with drooling of saliva (sialorrhoea), provide advice on swallowing, diet, posture, positioning, oral care and suctioning. **[2016]**

1.8.12 Consider a trial of antimuscarinic medicine as the first-line treatment for sialorrhoea in people with MND.

In February 2016 this was an off-label use of antimuscarinic medicine. See NICE's information on prescribing medicines. **[2016]**

1.8.13 Consider glycopyrrolate as the first-line treatment for sialorrhoea in people with MND who have cognitive impairment, because it has fewer central nervous system side effects.

In February 2016 this was an off-label use of glycopyrrolate. See NICE's information on prescribing medicines. **[2016]**

1.8.14 If first-line treatment for sialorrhoea is not effective, not tolerated or contraindicated, consider referral to a specialist service for Botulinum toxin A. **[2016]**

In September 2019 Xeomin was the only Botulinium toxin A with a UK marketing authorisation for this indication.

1.8.15 If a person with MND has thick, tenacious saliva:

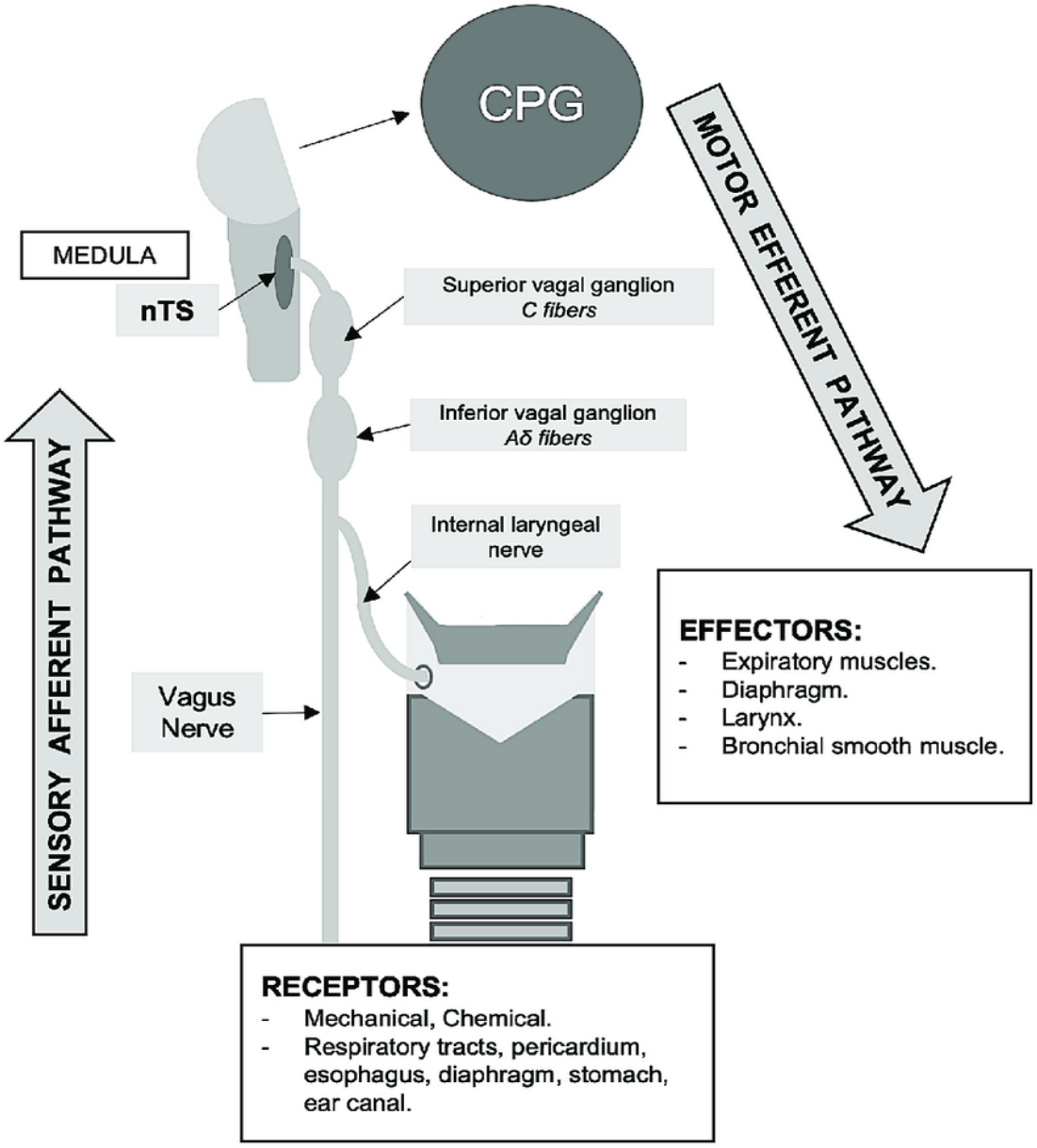
- review all current medicines, especially any treatments for sialorrhoea
- provide advice on swallowing, diet, posture, positioning, oral care, suctioning and hydration
- consider treatment with humidification, nebulisers and carbocisteine. **[2016]**

Thick tenacious chest secretions

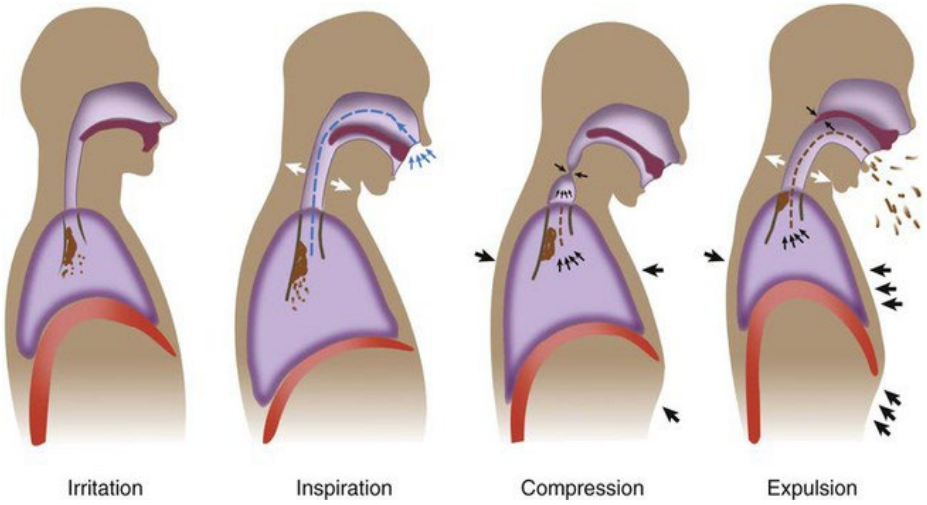
- Thinning secretions aids chest clearance.
- Consider:
  - Hydration status
  - Carbocysteine
  - Saline nebulisers
  - Underlying lung disease such as COPD/ bronchiectasis consider LT antibiotics such as Azithromycin/ N-acetylcysteine.

Chest secretion management

- Reduced cough function predispose to chest infection. Inability to clear chest secretions. Natural defence mechanism against aspiration of particles into the lung.


CPG
MOTOR EFFERENT PATHWAY
MEDULA
nTS
Superior vagal ganglion
C fibers
Inferior vagal ganglion
Aδ fibers
Internal laryngeal nerve
SENSORY AFFERENT PATHWAY
Vagus Nerve
EFFECTORS:
- Expiratory muscles.
- Diaphragm.
- Larynx.
- Bronchial smooth muscle.
RECEPTORS:
- Mechanical, Chemical.
- Respiratory tracts, pericardium, esophagus, diaphragm, stomach, ear canal.

Cough mechanism



Insufflation phase

Build up of intra-thoracic pressures

Expulsion phase

Cough impairment in ALS

- Bulbar dysfunction

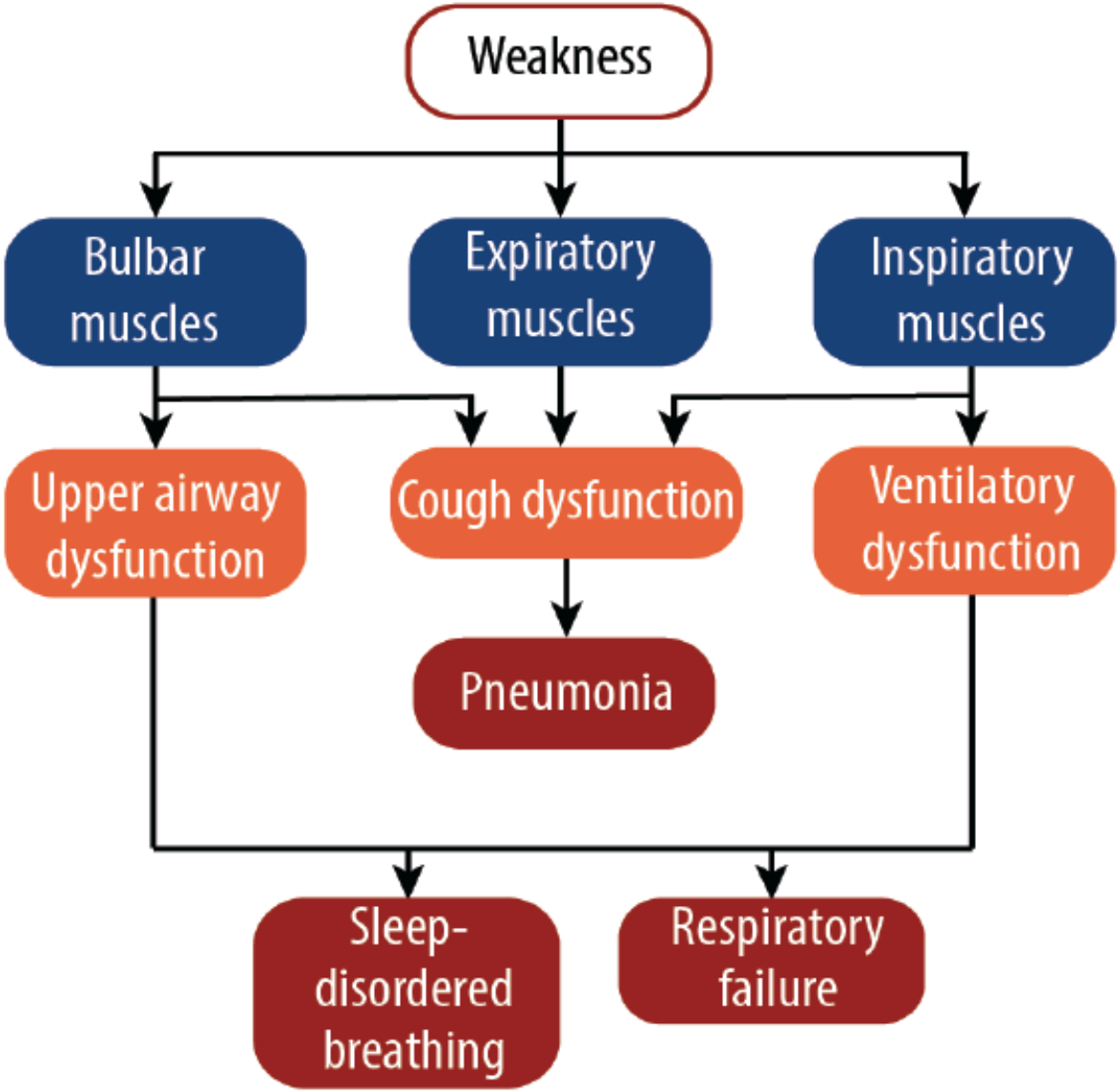
Reduces the rapid opening and closing of the glottis during coughing

- Inspiratory muscle weakness

Unable to reach maximal inspiratory capacity

- Abdominal/expiratory muscle weakness

Unable to generate adequate peak expiratory flow rates


Weakness
Bulbar muscles
Expiratory muscles
Inspiratory muscles
Upper airway dysfunction
Cough dysfunction
Ventilatory dysfunction
Pneumonia
Sleep-disordered breathing
Respiratory failure

Peak cough flow

360L/min

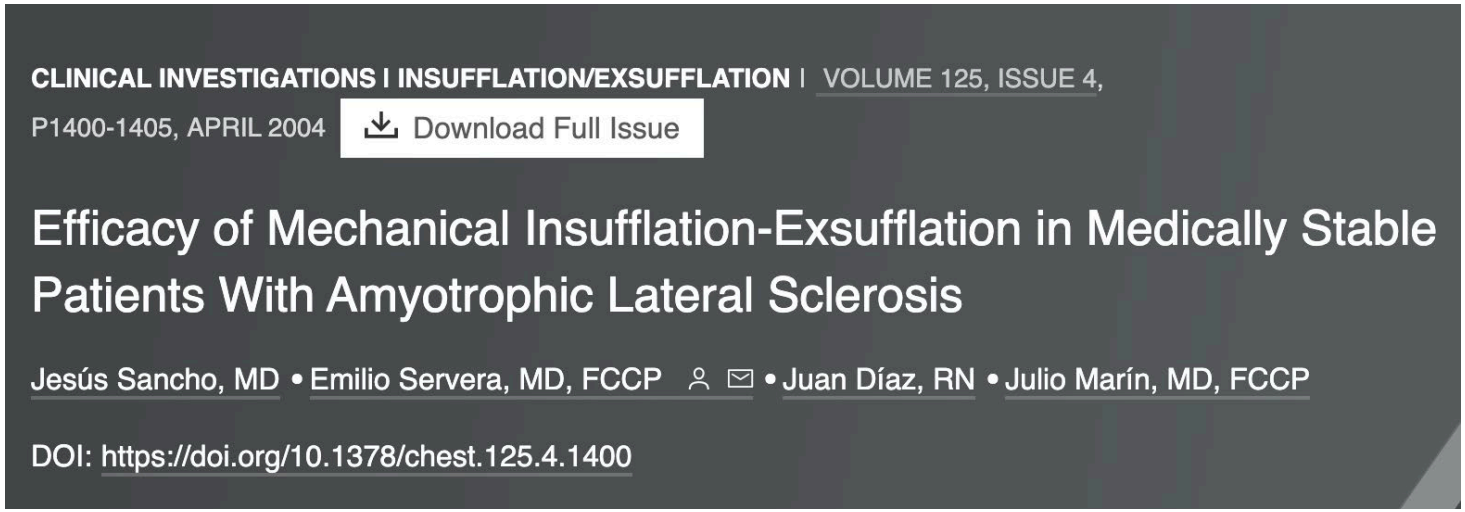
<360 >160L/ min

160L/min

Cough augmentation strategies

Treatment modality	Phase of cough augmentation	Ease of delivery	Additional benefits
Lung volume recruitment bag (LVR, breath-stacking)	Insufflation phase (1)	Can be self administered if UL strength adequate Can teach caregiver to apply	• Helps to maintain chest wall compliance • Portable • Not battery operated
Manually assisted cough (MAC)	Expulsion phase (3)	Cannot be applied by individual	
Mechanical insufflation-exsufflation (MI:E, Cough assist)	Insufflation phase (1) Expulsion phase (3)	Can be self administered if UL strength adequate Can teach caregiver to apply	• Acutely enhanced forced vital capacity • Helps maintain chest wall compliance • May avoid need for emergency help

Evidence base



CLINICAL INVESTIGATIONS I INSUFFLATION/EXSUFFLATION I VOLUME 125, ISSUE 4,
P1400-1405, APRIL 2004 Download Full Issue

Efficacy of Mechanical Insufflation-Exsufflation in Medically Stable Patients With Amyotrophic Lateral Sclerosis

Jesús Sancho, MD • Emilio Servera, MD, FCCP • Juan Díaz, RN • Julio Marín, MD, FCCP

DOI: https://doi.org/10.1378/chest.125.4.1400

- N = 26 PwALS (15 with severe bulbar dysfunction
- Numerous lung capacity/volume outcome measures:

Results:

- All pts had similar time from disease onset to diagnosis
- Statistical differences were found between the non-bulbar & bulbar patients in lung function and cough capacity parameters
- Four patients with bulbar dysfunction and MIC >1L had $PCF_{MI:E}$ < 2.7L/s

A preliminary randomized trial of the mechanica insufflator–exsufflator versus breath–stacking technique in patients with amyotrophic lateral sclerosis

Muhammad K Rafiq [1], Michael Bradburn [2], Alison R Proctor [1], Catherine G Billings [3], Stephen Bianchi [3], Christopher J McDermott [1], Pamela J Shaw [1]

- RCT
- 40 patients randomized to breath-stacking technique (n=21), MI:E (n=19)
- Patients followed up at 3-month intervals for at least 1yr or until death
- All patients diagnosed with respiratory failure and offered NIV

- Primary outcome measure:

No. of days with symptoms of chest infection, treated with antibiotics, in the community or in hospital.

- Secondary outcome measures: Survival and QoL benefit

A comparison of assisted cough techniques in stable patients with severe respiratory insufficiency due to amyotrophic lateral sclerosis

Cristina Senent, Jean-Louis Golmard, François Salachas, Eusebi Chiner, Capucine Morelot-Panzini, Vincent Meninger, ...

Pages 26-32 | Received 26 Apr 2010, Accepted 21 Oct 2010, Published online: 24 Nov 2010

- Unassisted PCF vs $PCF_{MI:E}$

- Bulbar group, these values were 42 (35–130) l/min versus 436 (244–630) l/min, respectively (p = 0.008), and 89 (40–106) l/min versus 491 (192–580) l/min, respectively, in the non-bulbar group (p = 0.019)

The effects of lung volume recruitment on coughing and pulmonary function in patients with ALS

Stuart Cleary, John E. Misiaszek, Sanjay Kalra, Sonya Wheeler & Wendy Johnston

Pages 111-115 | Received 10 May 2012, Accepted 05 Aug 2012, Published online: 12 Sep 2012

LVR had a significantly positive effect on FVC for up to 15 min following treatment but did not have a facilitative effect on SnP at any time-point.

LVR had a significantly positive effect on PCF during unassisted coughing at both 15 min and 30 min following treatment, and there was no significant decrease in flow rates from baseline to 30 min later.

Observational Study > Thorax. 2017 Mar;72(3):221-229. doi: 10.1136/thoraxjnl-2015-207555. Epub 2016 May 12.

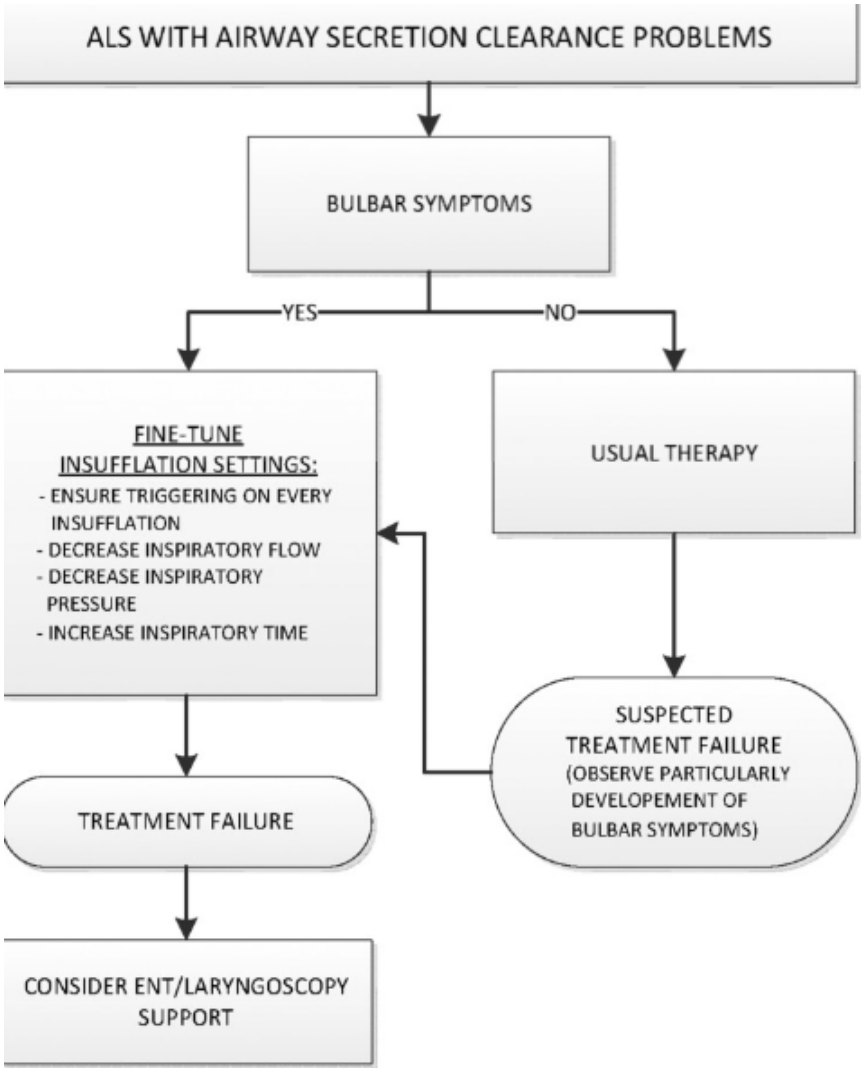
Laryngeal response patterns influence the efficacy of mechanical assisted cough in amyotrophic lateral sclerosis

Tiina Andersen [1 2 3], Astrid Sandnes [3], Anne Kristine Brekka [4], Magnus Hilland [5], Hege Clemm [3 6], Ove Fondenes [1], Ole-Bjørn Tysnes [7 8], John-Helge Heimdal [5 8], Thomas Halvorsen [3 6], Maria Vollsæter [1 3 6], Ola Drange Røksund [4 6]

- Cross-sectional study of 20 patients with ALS and 20 healthy age-matched and sex-matched volunteers
- Video-recorded flexible transnasal fibre-optic laryngoscopy
- Standardised protocol, applying pressures of ±20 to ±50 cm H_2O
- Laryngeal movements were assessed from video files

Results

- At the supraglottic level, all patients with ALS and bulbar symptoms (n=14) adducted their laryngeal structures during insufflation
- At the glottic level, initial abduction followed by subsequent adduction was observed in all patients with ALS during insufflation and exsufflation
- Hypopharyngeal constriction during exsufflation was observed in all subjects, most prominently in patients with ALS and bulbar symptoms
- Healthy subjects and patients with ALS and no bulbar symptoms (n=6) coordinated their cough well during MI-E



Andersen et al., 2017

The experiences and perceptions of mechanical insufflation-exsufflation in people with amyot lateral sclerosis

Shauna Sheridan, Joerg Steier, Philip Marino, Shelley Srivastava, Eui-Sik Suh, Nicholas Hart, Patrick Murphy, Harri Leyla Osman, Georgios Kaltsakas, Michelle Ramsay

Lane Fox Respiratory Service, Guy's and St Thomas' NHS Foundation Trust, London.

Introduction

- Amyotrophic lateral sclerosis (ALS) is an incurable progressive neuromuscular disease.
- Focus of treatments are to alleviate symptoms, thereby improving quality of life.
- Mechanical in-exsufflation (MI:E) is a respiratory adjunct used to improve cough function and enhance secretion clearance from the airways.
- Limited research is available on the experiences and perceptions of MI:E in people with ALS (pwALS).
- A service evaluation was carried out to investigate this.

Aims & Objectives

Aims:

- To explore pwALS experiences and perceptions of using MI:E in the home setting

Objectives:

To conduct semi-structured interviews to:

- To understand patients' experiences and perceptions of MI:E
- To understand the impact of MI:E on patients' quality of life
- To identify facilitators and barriers to MI:E usage

Figure 1. Flow diagram of the recruitment process

Figure 2. Interview types
F2F = Face to face

'Maybe checking in every month or something to see how it's going along. So, it makes you feel guilted into using it'

'I think it takes Ruth several minutes to set it all up and its really to eh, safeguard her as much as anything'

Methods

- PwALS meeting the inclusion criteria were invit participate in semi structured interviews.
- Interviews were transcribed and analysed using the analysis.
- Coding was performed using qualitative data a software, Nvivo 12

Results

- Perception of MI:E is not fixed
- Factors that impact on MI:E efficacy
- The impact of MI:E is multi-faceted
- Facilitators to usage
- Barriers to usage

Figure 3. The five overarching themes

Conclusion

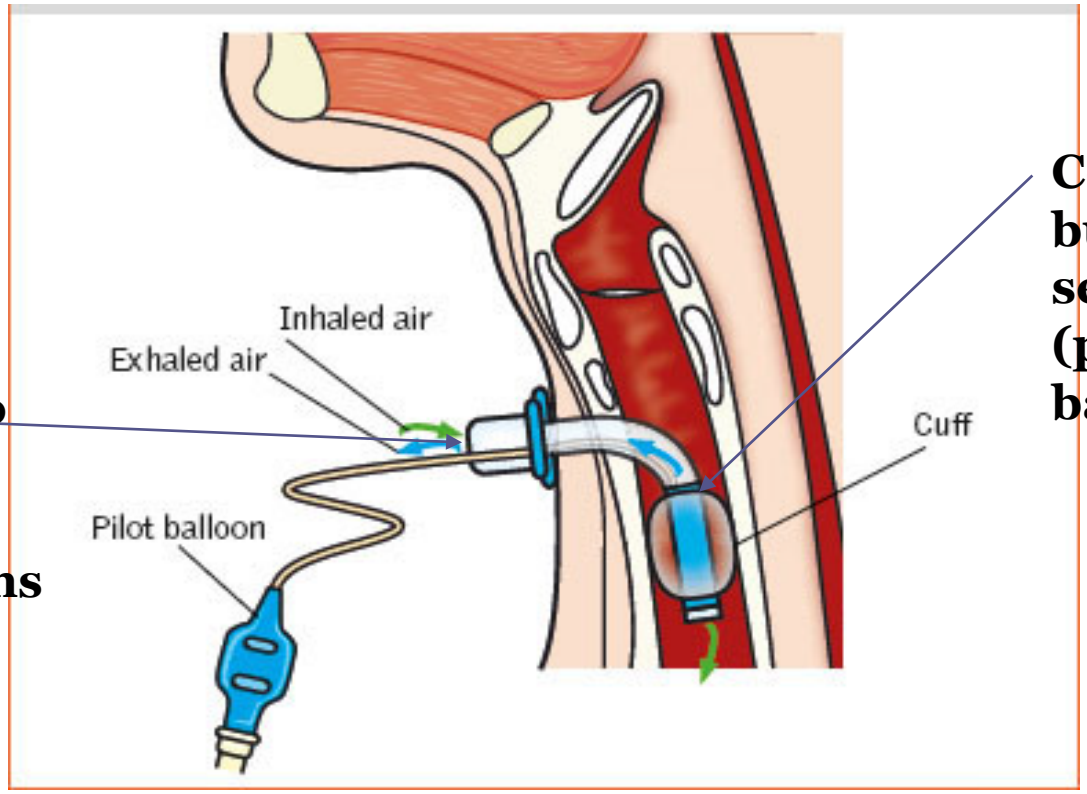
- Lived experience of pwALS using MI:E is not fixed, cha from fear and grief to physical relief and reassurance
- Ensuring adequate training of the operator is essenti enhance MI:E effectiveness and pwALS experience
- Regular review by the issuer to ensure that patient and caregivers receive adequate support concerning MI:E usag

My personal view

- Holistic approach to cough assessment
- Peak cough flow measurement is only one part of the overall assessment
- Patients with severe bulbar dysfunction require careful consideration
- Does prophylactic treatment increase burden of care?

Tracheostomy

Direct access to suction chest secretions



Control of bulbar secretions (physical barrier)

Role tracheostomy ventilation (paucity of data)

Advantages

- May improve ventilation in appropriate patients
  - due to reduction of leak
  - increased ventilatory security
  - Bypass upper airway issues- sialorrhea, glottic closure
- Liberates face
  - Enhances interaction with relatives
  - Facilitates oral hygiene/tastes for pleasure in ventilator dependent patient
- Enhanced secretion management
  - Facilitates direct suction of chest secretions
- Case reports of improved survival

Difficulties

- Risk of procedure and GA
  - Mortality reported as high as 10%
- Potential loss of voice/swallow
- Increased support required
  - will need 24hr care with tracheostomy trained carers
  - Impact on independence/intimacy with partners/**carer burden**
- Significant delays in discharge
  - CHC/ICB funding
  - Lack of trained carers/ nurses
- High associated costs (>250k/yr)
- May survive with increased levels of disability

Many thanks for your attention.

We would be happy to take your questions.

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