Key points

- Ineffective cough is a major cause of mortality and morbidity in patients with neuromuscular disease.
- A normal cough requires the inspiratory muscles to inspire to up to 85–90% of total lung capacity followed by rapid closure of the glottis for ~0.2 s. Both glottic opening and contraction of abdominal and intercostal (expiratory) muscles, resulting in intrapleural pressures of >190 cmH_2O and generating transient peak cough flows (PCF) of 360–1,200 L per min [1] complete the manoeuvre.
- Cough assist techniques are used in patients who present with a weak cough. The goal of these techniques is to increase the expiratory airflow that occurs during a cough, by assisting inspiration and/or expiration, thus increasing cough efficacy.
- When conventional cough assistance techniques become ineffective, a mechanical insufflator–exsufflator should be considered.

The ERS designates this educational activity for a maximum of 1 CME credit. For information on how to earn CME credits, see page 375

How to use a mechanical insufflator–exsufflator "cough assist machine"

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Educational aims

- To raise awareness of how to use mechanical insufflator–exsufflators.
- To discuss the use of mechanical insufflation–exsufflation compared with conventional airway clearance techniques.
- To identify an escalation treatment protocol.

Summary

Effective cough is a protective mechanism against respiratory tract infections. In patients with respiratory muscle weakness due to neuromuscular disease, respiratory tract infections are the commonest cause of hospital admission [2]. Neuromuscular disease patients may have impaired cough and a reduction in PCF [3] as a result of inspiratory and expiratory muscle weakness, which causes a reduction in the pressure available to drive the cough manoeuvre [4]. While techniques such as breath stacking and manually assisted coughing can improve cough strength, these eventually become ineffective in some adult and paediatric patients.

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Provenance
Commissioned article, peer reviewed

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 simulating the airflow changes that occur during a cough, thereby assisting sputum clearance.

In 1953, various portable devices were manufactured for the delivery of mechanical insufflation–exsufflation (MI–E; *e.g.* Cofflator portable cough machine; OEM, St Louis, MO, USA). The most commonly used mechanical insufflator–exsufflators today are the Cough Assist (JH Emerson Co., Cambridge, MA, USA; figure 1) and the Pegaso (Dima Italia, Bologna, Italy). Initial investigations showed MI–E to be effective at removing foreign bodies from anaesthetised dogs [5]. BECK and BARACH [6] demonstrated clinical and radiographic improvement in 92 out of 103 acutely ill patients with respiratory tract infections with the use of MI–E, in a study that included 72 patients with lung disease and 27 with skeletal or neuromuscular disease. Greater improvements were seen in patients with neuromuscular disease. There have been no reports of significant barotrauma or pneumothorax. The cardiovascular effects of MI–E were evaluated by BECK and SCARRONE [7], who found that patients demonstrated an increase in mean heart rate by 17 beats per min and an increase in systolic blood pressure of 8 mmHg, but no major adverse effects. This latter study also found an increase in cardiac output by 2.1 L per min and echocardiograph changes reflective of rotation of the heart during normal coughing.

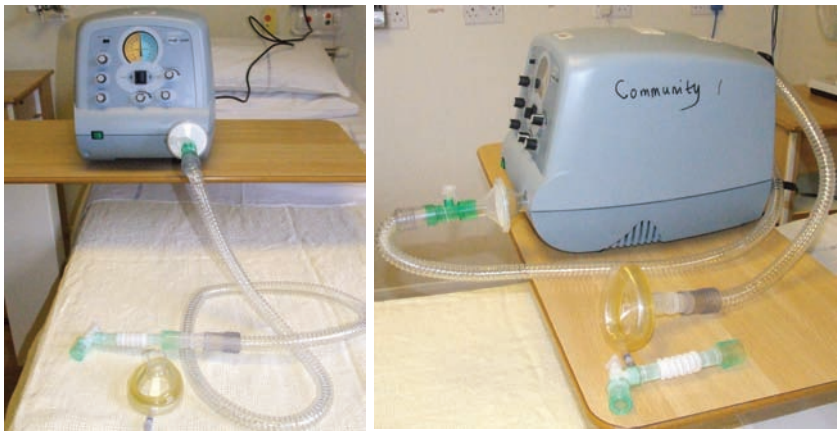


Figure 1
Mechanical in–exsufflator (Cough Assist) manufactured by JH Emerson Co. Circuit consists of antibacterial/antiviral filter, wide-bore tubing, male:male connector (22 mm) and either a catheter mount or mask.

Conditions that may benefit from MI-E

Physiologically, MI–E has been shown to increase peak cough flow (PCF) in patients with neuromuscular disease [3, 4, 8–10]; an increase in PCF is thought to improve the efficacy of the cough and thus assist in secretion removal. MUSTFA *et al.* [4] and SANCHO *et al.* [9] found a significant improvement from baseline PCF with MI–E for both bulbar and nonbulbar motor neurone disease patients. MI–E has not been shown to be effective in patients with chronic obstructive pulmonary disease [10, 11]. Theoretically, however, it might benefit any individual in whom ventilatory capacity is outstripped by ventilatory load, *e.g.* weaning patients.

Guide to setting up MI–E

The device can be used in manual or automatic mode. Initially, the device is set up in manual mode with a full face mask; this ensures maximum comfort for the patient. Often the device is then changed to automatic mode, enabling it to be used in the domiciliary environment without a trained professional. Initially, oxygen saturation should be monitored, especially during an acute chest infection. See figure 2 for the basic controls of the Cough Assist and Pegaso units. The box gives an introduction to setting up the Cough Assist machine.

- Start the insufflation (positive) pressure at 15–20 cmH_2O (1.5–2.0 kPa) and increase to give an inspiration to total lung capacity. You may require a positive pressure as high as 40 cmH_2O (3.9 kPa; pressure delivered is often limited by the mask lifting off the face at high levels). Insufflation should last ~2 s, or longer if required and should be titrated for patient comfort.
- Start with the exsufflation (negative) pressure the same as the insufflation pressure, then increase the negative pressure to 10–20 cmH_2O (1.0–2.0 kPa) above the positive pressure.The negative pressure is then held for 3–6 s, simulating the airflows that occur naturally during the cough. The best indicator of efficacy is an increase in the sound of the cough: listen carefully. The patient is often able to gauge efficacy well.
- The patient will learn to coordinate their cough when the device switches to exsufflation. Inform the patient a deep breath is coming and tell them to cough as they feel the negative pressure.
- Alternatively, for the Pegaso mechanical in–exsufflator settings are input *via* a menu. Insufflation is then set by either increasing or decreasing the up or down setting. The unit works either in an automatic mode (setting insufflation time, exsufflation time and pause) or in manual mode by pressing "I" or "E" (insufflation or exsufflation). Both devices use the same circuit set-up (filter-tubing-mask, as shown in figure 1).

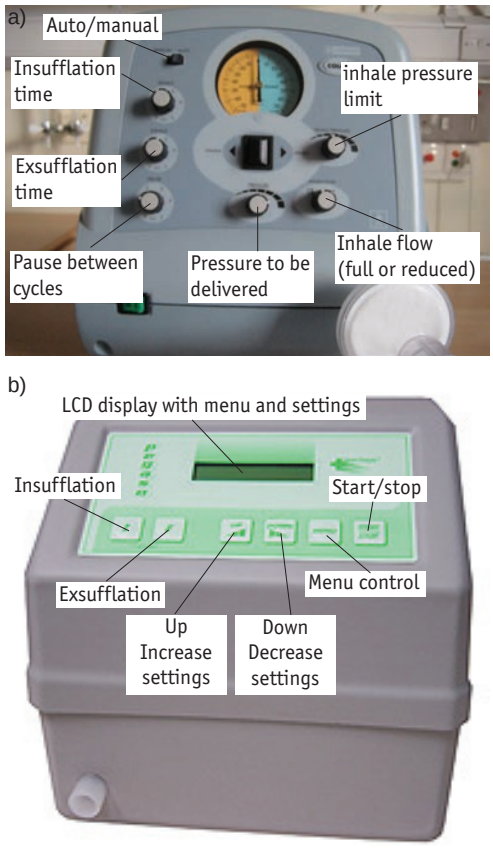


Figure 2
What the buttons do on a) the Cough Assist machine and b) the Pegaso mechanical in-exsufflator.

Examples of settings used in studies

Various authors [3, 10, 12, 13] have reported a good outcome with low pressures. One study in a predominant paediatric population (age range 3 months–28.6 years) used median pressures of +30 to -30 cmH_2O (+2.9 to -2.9 kPa), with a range of +15 to +40 cmH_2O (+1.5 to +3.9 kPa) for insufflation and -20 to -50 cmH_2O (-2.0 to -4.9 kPa) for exsufflation [12]. However, an alternative centre [14–19] advocates higher pressure spans (+40 to -40– -60 cmH_2O; +3.0 to -3.9– -5.9 kPa).

Use of MI–E in the acute setting

VIANELLO *et al*. [13] compared 11 neuromuscular disease patients who received a mean±SD 2.7±0.9 MI–E sessions per day to 16 historical matched controls who were treated with postural drainage, manually assisted coughing and suction when required. The results of the retrospective study showed that patients who received MI–E had a lower treatment failure rate (defined by the need to insert a mini-tracheostomy or the need to intubate; 18 *versus* 62.5%; p=0.047). There was no significant difference between the groups in the proportion of patients requiring noninvasive positive pressure ventilation (NIPPV; p=0.37) or *i.v.* antibiotics (p=0.25), in the duration of NIPPV (p=0.93) or in the proportion of patients requiring bronchoscopy (p=0.71).

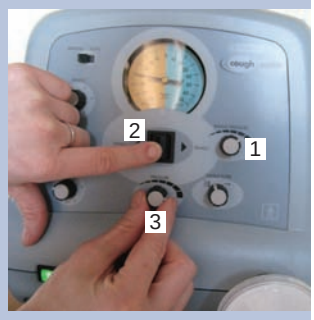
The present author's group studied eight patients (six males) with neuromuscular disease with a median (range) age 21.5 (4–44) years. Diagnoses included Duchenne muscular dystrophy and spinal muscular atrophy. All had difficulty clearing secretions and a symptomatic acute chest infection. Patients underwent a 2-day randomised treatment programme of a modified active cycle of breathing techniques with increased NIPPV settings (P+NIPPV) and assisted coughing for one session and the same treatment but with the addition of MI–E for a second session, with crossover the next day. No difference in mean heart rate, oxygen saturation or transcutaneous measurement of carbon dioxide was found during either treatment. There was a significant improvement in air entry for both treatment sessions. Treatment time after 30 min was significantly shorter in the sessions that had the addition of MI–E (median (range) 17 (0–35) min *versus* 0 (0–26) min; p=0.03). Patients reported subjectively better visual analogue scores for the amount of sputum cleared in the treatment session with the addition of MI–E. The present author's group concluded that, from a respiratory physiotherapy point of view, MI–E procedures are an extremely useful addition to the assisted coughing toolbox. A similar effectiveness in airway clearance was seen with conventional physiotherapy but the treatment time needed to clear secretions was longer. However, it is important to remember that one should try simpler and less costly methods of cough assistance (*e.g.* breath stacking and manually assisted coughs) prior to the use of MI–E, if time permits.

Setting up the Cough Assist

Setting the pressure

Block off the end of the tubing.

1. *Set the inhale pressure to full.*
2. *Push the manual control lever to exhale.*
3. *Turn the pressure button until the desired exsufflation pressure is reached.*



Setting the inhale pressure lower than the exhale pressure

Block off the end of the tubing.

1. *Push the manual control lever to inhale.*
2. *Turn the inhale pressure button until the desired insufflation pressure is reached.*

Setting up automatic mode

Set the device as previously described.

1. *Set the inhale time to the desired time in s.*
2. *Set the exhale time to the desired time in s.*
3. *Set the pause time to the desired time in s (i.e. the amount of time to rest between insufflation and exsufflation cycles).*
4. *Set the switch to automatic.*

Setting inhale flow full or reduced

1. *The inhale flow button allows a decreased inhale pressure to be delivered.*

Educational questions

Are the following statements true or false?

1. Cough in–exsufflation is useful in the acute consolidation phase of pneumonia.
2. Bulbar patients do not benefit from MI–E.
3. MI–E has been shown to increase peak cough flow in neuromuscular patients.
4. MI–E should be used in the absence of secretions.
5. You should always use the highest insufflation–exsufflation pressures, regardless of patient comfort.

Frequently asked questions

Can I use MI-E in an infant?
MI-E can be used in small children. We have clinical experience in children as young as 3 months old. One needs to be aware that infants have limited collateral ventilation and are nearer to their physiological closing volume; consequently you should start off with lower pressures. Using MI-E does not necessarily negate the need for nasopharyngeal suction to clear secretions when initially using the technique.

Can I use MI-E with a mouthpiece?
MI-E can be delivered *via* a mouthpiece. However, when the patient coughs and opens the mouth a good seal may be lost.

What are the correct settings?
There is no one setting for all, nor a simple algorithm. Everyone is an individual and some patients may require higher settings than others. Ask the patient to cough into the circuit and remember the quality of the cough. Start the pressures low (+15 to +20, -20 to -30 cmH_2O; +1.5 to +2.0, -2.0 to -2.9 kPa) on the MI-E and build up to a good chest expansion; adjust the negative pressure until the audible quality of the cough has improved.

Can MI-E be used in patients with bulbar problems?
MI-E is effective in individuals with bulbar weakness. However, if the bulbar weakness is very severe, MI-E may be insufficient so a tracheostomy should be considered, if appropriate.

Can patients with learning disabilities use MI-E?
MI-E should be attempted and can be used very effectively in patients with learning difficulties, although it may take longer to acclimatise them to the device. The device is used in an attempt to stimulate a natural cough. The best coughs are achieved with coordination of a cough at the point of exsufflation.

How many times a day should MI-E be used?
MI-E can be used as many times during the day as is required to clear secretions. It is important to remember that lots of coughing does induce fatigue, so patients should be allowed adequate rest periods. If you need to use MI-E frequently, then you should consider whether a long enough airway clearance session has been carried out in the first place and amend treatment duration accordingly. Are the secretions due to drooling? If so, consider hyoscine patches or glycopyrrolate.

How long should treatment sessions with MI-E be continued?
At our centre, we have found a typical airway clearance session in a neuromuscular disease patient can last 30–60 min during an acute severe chest infection. Often, treatments last until the patient is clear of secretions or until they are tired and require a rest.

Should MI-E be used if there are no secretions present?
If there is an area of collapse and/or consolidation on the chest radiograph and the patient has an ineffective cough it is worth trying. Some patients who have the device at home use it daily to ensure there are no secretions present. Additionally, some patients who have the device at home use it daily to provide a stretch to the chest wall. If there are no secretions present and there is no other indication to use MI-E, then daily sessions do not need to be carried out.

Can MI-E be used if the patient has had a pneumothorax?
If a patient has an undrained pneumothorax, MI-E is contraindicated. If the patient has had a pneumothorax in the past that has resolved, MI-E can be used with caution. If the patient has pneumothorax with a drain *in situ*, a clinical decision should be made, balancing risk against benefit in that particular case, as negative and positive pressure may aggravate air leak.

What are the contraindications to MI-E?
MI-E is not advisable in patients with uncontrolled asthma or bronchospasm. It is worth noting that some wheezy airway sounds may be generated by secretions, so all clinical circumstances should be taken into consideration. MI-E should not be used in hypotensive patients or those with significant haemoptysis.

Can the same mechanical insufflator–exsufflator be used with different patients?
The device will need to be cleaned between patients and each patient should have their own circuit; settings should be checked prior to commencement of treatment. A high-efficiency bacterial filter should be placed between patient circuit and device. The device should not be taken from dirty to clean areas as defined by local infection control policy.

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Suggested answers

1. False.
2. False.
3. True.
4. False.
5. False.

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Commentary: Exploring the potential of mechanical insufflation–exsufflation

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Provenance
Commissioned editorial comment

Cough augmentation with mechanical insufflation–exsufflation (MI–E) has been described as a technique that facilitates airway secretion clearance in different neuromuscular disorders (NMD), thus avoiding hospitalisations, and preventing pneumonias and episodes of respiratory failure for patients with Duchenne muscular dystrophy [1, 2] spinal muscular atrophy [3], high spinal cord injury [4] and amyotrophic lateral sclerosis (ALS) [5]. Although there are a number of guidelines and consensus statements that emphasise its importance [6–9], MI–E is far from widely used [10, 11].

In the present issue of *Breathe*, M. Chatwin describes a very practical guide to the application and principal indications of MI–E for different settings that may encourage its use; however, some important considerations should be discussed to explore the full potential of this technique.

The importance of the use of manually assisted coughing for the optimisation of peak cough flows (PCF) has been demonstrated [12, 13] but it cannot be a comparable alternative to MI–E, rather an MI–E complement; so, except after a meal, applying MI–E with an abdominal thrust in conjunction with the exsufflations must be taken into consideration in order to optimise the technique [14].

Insufflation and exsufflation pressures

A normal cough expels a volume of air four times greater than a normal tidal volume. Therefore, provision of a normal tidal volume for NMD patients whose tidal volumes approach their vital capacities will not optimise their cough flows; this can never be accomplished for these patients by limiting insufflation and exsufflation pressures to 25 and -25 cmH_2O (2.4 and -2.4 kPa) [15].

For patients with NMD undergoing MI–E, barotrauma is rare. In >1,000 ventilator users, most of whom were dependent on MI–E with insufflation pressures of 40–60 cmH_2O (3.9–5.9 kPa) to spare them from upper respiratory tract infections (in some cases over a 52-year period), no pneumothoraces were ever found [16, 17].

The maximal effectiveness of MI–E at pressures of 40 to -40 cmH_2O (3.9 to -3.9 kPa) has been demonstrated in experimental models [15, 18] and both in adult [19] and paediatric populations [20]. Although MI–E pressures of 40 to -40 cmH_2O (3.9 to -3.9 kPa) are generally adequate for most patients, higher settings are often required when compliance decreases (by obesity or scoliosis) or resistance increases. In fact, SANCHO *et al*. [15], in a lung model, demonstrated the need for 70 to -70 cmH_2O (6.8 to -6.8 kPa) in order to achieve a clinically effective PCF of 2.67 L per s.

When using MI–E *via* small translaryngeal or tracheostomy tubes, higher pressures of 60–70 cmH_2O (5.9–6.8 kPa) is recommended to overcome the tube resistance, and cuffs should be inflated to prevent leaks. In this context, it has been demonstrated that MI–E *via* a tracheostomy tube is more effective in clearing secretions than conventional suctioning, and it is preferred by patients [21, 22].

Time settings

Although the machine can be managed automatically by programming the insufflation/exsufflation/pause times, the manual mode permits a better synchronisation and it is easier for patients to coordinate their insufflation and cough with the machine. This is especially true for infants with spinal muscular atrophy type 1, in whom the cycles should follow their rapid respiratory rate and chest movement [3]. Insufflation and exsufflation times should be adjusted to provide maximum chest expansion and lung emptying, respectively. If insufflation or exsufflation pressures and times are inadequate, the patient literally cannot breathe.

The application of MI–E

MI–E has been also described as a very efficient technique in the acute setting for NMD patients, in the treatment of respiratory failure due to upper respiratory tract infections [23], to avoid intubation [24], to facilitate extubation and decannulation and to prevent post-extubation failure [25–27]. However, the evidence supporting the role of this technique to facilitate extubation in difficult weaning patients is lacking. Our group has started to study how this technique can improve the efficacy of noninvasive ventilation in these patients, and the results so far are promising.

The application of MI–E in the post-operative phase has been also described both in initial and recent papers [28, 29]. In this context, as experience with MI–E increased, bronchoscopy was performed less frequently for the removal of bronchial secretions in our centre. For example, in an 18-year-old male with myotonic dystrophy who developed left lung atelectasis and respiratory failure after Nuss operation (for pectus excavatum), intensive MI–E avoided endotracheal intubation and bronchoscopy (figure 1).

Both in the acute and chronic setting, MI–E is labour intensive and often difficult for non-professional caregivers. However, it is next to impossible to manage advanced NMD patients without tracheostomy tubes, unless their families and care providers provide virtually all of their care during upper respiratory tract infection. It is inadequate for hospital staff to take full care of the hospitalised patient and instruct the family just before discharge, and then to expect another episode to be prevented. Whereas the family often has the time and motivation to use MI-E along with abdominal thrusts, sometimes every 15 min, and to use oximetry as feedback to maintain normal saturation (without supplemental oxygen) for the home or intensive care patient, one cannot expect the respiratory physiotherapy and nursing staff to do this.

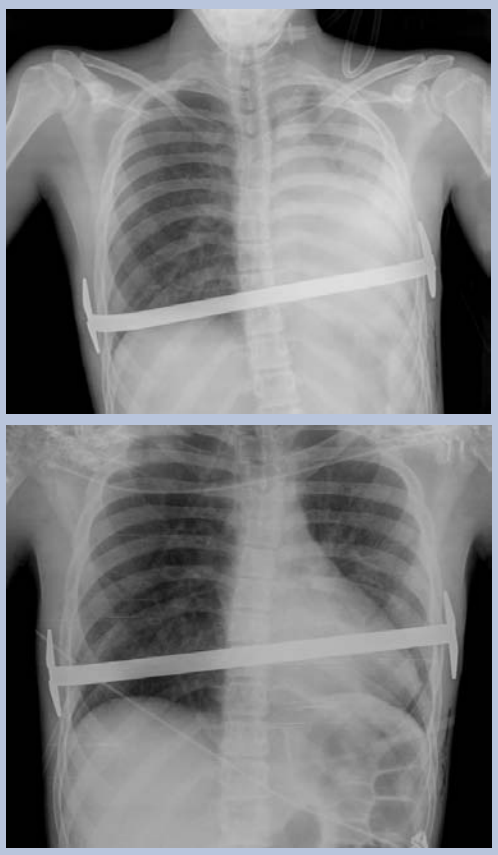

Figure 1
Eight sessions of MI-E applied to a patient with myotonic distrophy (following surgical procedure and general anaesthesia) reversed respiratory failure and atelectasis. Vital capacity, PCF and oxygen saturation improved with clearing of mucous plugs (vital capacity from 0.69 to 1.71 L, PCF from 175 to 350 L per min and oxygen saturation measured by pulse oximeter from 88 to 97%).

In NMD patients, MI–E failure only occurs in those who cannot cooperate (unless they have an endotracheal tube) or who have severe bulbar dysfunction [30]. In some bulbar ALS patients, MI–E can induce upper airway collapse, making the technique uncomfortable and ineffective [15]. In fact, the current authors have had two ALS patients (one with a floppy epiglottis and one with severe tongue weakness), in whom MI–E (and also nasal ventilation) caused stridor due to epiglottic prolapse and glossoptosis (figure 2). However, even in bulbar ALS it worthwhile attempting MI–E, since there are some patients that can improve PCF [31] and are able remove secretions during a chest infection [32].

According to the present authors' experience, MI–E can be very effective in NMD patients who are on continuous noninvasive ventilatory support for years despite no ventilator-free breathing ability, and it is possible to apply this technique proactively with oximetry feedback to

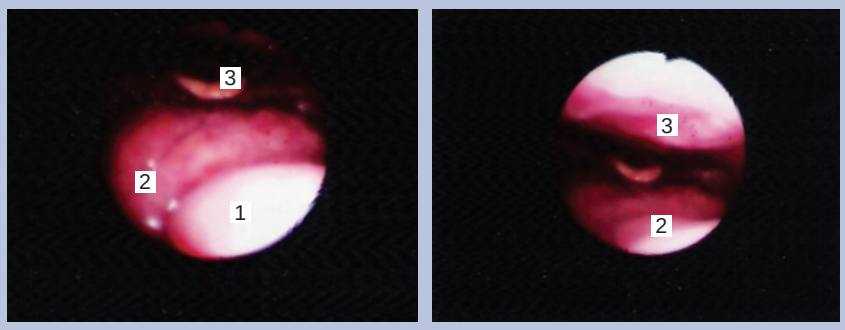

Figure 2
Nasopharyngoscopy: glossoptosis during positive pressure in a severe bulbar ALS patient with tongue weakness. 1: uvula; 2: tongue; 3: epiglottis.

avoid patients becoming hospitalised or developing pneumonia.

Conclusion

MI-E is very effective in the resolution of acute respiratory failure in NMD patients, but is rarely needed for stable patients with intact bulbar function who can air stack to maximum lung volumes [33] and close the glottis against high pressures with an abdominal thrust. However, even in stable patients it may be advisable to use it routinely, just to stay in practice so that the patient can apply it in an effective way during upper respiratory tract infections (as that is the time when the technique is most needed).

In conclusion, the use of MI–E can be time consuming but, without doubt, avoids hospital admissions and fibreoptic bronchoscopy for atelectasis, and reduces hospital stay, thus contributing to an improvement in patients' quality of life and a reduction of hospital costs. Further exploration of the use of this technique in the acute setting is thus warranted.

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