



Original Article

Indications and Compliance of Home Mechanical Insufflation-Exsufflation in Patients with Neuromuscular Diseases

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ABSTRACT

Introduction: Neuromuscular disease (NMD) patients frequently have impaired cough. Mechanical insufflation-exsufflation (MI-E) has proven efficacy in improving airway clearance, however data related to its long-term home use is lacking. The purpose of this study was to describe indications, safety and compliance of home MI-E in NMD patients.

Methods: Four years observational analysis of 21 NMD patients on home MI-E. Diagnosis included bulbar and non-bulbar Amyotrophic Lateral Sclerosis (ALS) and other NMD. Median age was 58 years. Only cooperative patients with unassisted baseline Peak Cough Flow (PCF) < 270 L/min were included. All patients were under continuous mechanical ventilation (6 by tracheostomy). Pulmonary function before initiation of MI-E (median): FVC = 0.81 L, MIP = 28 cmH₂O, MEP = 22 cmH₂O and PCF = 60 L/min. MI-E was performed by previously trained non-professional caregivers, with an on-call support of a trained health care professional. Patients had pulse oximetry monitorization and applied MI-E whenever SpO₂ < 95%. Median follow-up was 12 months (3-41 months).

Results: Ten patients (9 ALS) used MI-E daily. Eleven patients used MI-E intermittently, during exacerbations, and in 8 patients early application of MI-E (guided by oximetry feed-back) avoided hospitalization. All tracheostomized patients used MI-E daily and more times a day than patients under NIV. Four patients (3 bulbar ALS), were hospitalized due to secretion encumbrance. MI-E was well-tolerated and there were no complications. In general, caregivers considered MI-E effective. During this period, 4 patients died, related to disease progression.

Conclusions: Home MI-E is well tolerated, effective and safe if used by well trained caregivers. MI-E should be considered as a complement to mechanical ventilation.

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Indicaciones y cumplimiento con la insuflación-exuflación mecánica domiciliaria en pacientes con enfermedades neuromusculares

RESUMEN

Introducción: Con frecuencia, los pacientes con enfermedades neuromusculares (ENM) presentan un deterioro del mecanismo de la tos. Se ha demostrado la eficacia de la insuflación-exuflación mecánica (IEM) en la mejora del aclaramiento de las vías respiratorias aunque no se dispone de datos relacionados con su utilización domiciliaria a largo plazo. El objetivo del presente estudio fue describir las indicaciones, tolerabilidad y cumplimiento con la IEM domiciliaria en pacientes con ENM.

Palabras clave:

Insuflación-exuflación mecánica

Domicilio

Seguridad

Eficacia

Enfermedad neuromuscular

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Métodos: Análisis observacional de 4 años de duración de 21 pacientes ENM tratados con IEM domiciliaria. El diagnóstico incluyó esclerosis lateral amiotrófica (ELA) bulbar y no bulbar y otras ENM. La edad mediana fue de 58 años. Sólo se incluyeron pacientes cooperadores con valores de flujo pico de tos basal (FTM) no asistido < 270 l/min. Todos los pacientes estaban sometidos a ventilación mecánica continua (seis mediante traqueostomía). La función pulmonar previa al inicio de la IEM (mediana) era: FVC, 0,81 l, MIP, 28 cmH₂O, MEP = 22 cmH₂O y PCF = 60 l/min. Cuidadores no profesionales adiestrados previamente con el apoyo permanente de un profesional sanitario experto efectuaron la IEM. En los pacientes se monitorizó la pulsioximetría y se aplicó IEM siempre que la SpO₂ fue < 95%. El seguimiento mediano fue de 12 meses (3-41 meses).

Resultados: Utilizaron diariamente IEM 10 pacientes (nueve con ELA). La utilizaron de forma intermitente 11 pacientes durante las exacerbaciones y en 8 la aplicación precoz de IEM (guiada por la información de la oximetría) evitó la hospitalización. Todos los pacientes traqueostomizados utilizaron la IEM a diario y un mayor número de veces al día que los pacientes sometidos a ventilación mecánica no invasiva (VNI). Requiere ingreso hospitalario 4 pacientes (3 ELA bulbar) debido a la acumulación de secreciones. La IEM fue bien tolerada y no se asoció a complicaciones. En general, los cuidadores la consideraron eficaz. Durante este período, 4 pacientes fallecieron, en relación con la progresión de la enfermedad.

Conclusiones: La IEM domiciliaria es bien tolerada, eficaz y segura cuando la utilizan cuidadores adiestrados apropiadamente. Debe considerarse un complemento de la ventilación mecánica.

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Introduction

Patients with neuromuscular diseases (NMD) frequently have weak respiratory muscles and a deteriorated cough mechanism. Effective coughing depends on the capacity of the inspiratory muscles to achieve an inspiration of about 80% of total pulmonary capacity, followed by closure of the glottis and a pause with an increase in pulmonary volume.¹ After the contraction of the expiratory muscles, intrathoracic pressure increases and, when the glottis opens, air is expelled and secretions are propelled to the central airways.¹ Therefore, deterioration of the cough reflex in patients with NMD is related to weakness of the respiratory muscles, bulbar dysfunction that causes disability to control the glottis or even a deformity of the chest wall due to scoliosis.¹⁻⁵ Deterioration of the cough reflex causes insufficient airway clearance, pneumonia, atelectasia, and respiratory failure related to the accumulation of secretions.^{2,5} Based on basal function, a peak cough flow (PCF) < 160 l/min has been proposed as an indication of an ineffectual cough.^{2,5-8} However, Even a basal PCF < 270 l/min has been associated with pulmonary complications, since, during acute disease, there are additional reductions of the force of the respiratory muscles with an even greater reduction of PCF.^{2,9-11} As a result, management of airway secretions, especially during intercurrent airway infections, is a significant problem in patients with NMD and is the main cause of morbidity, prolonged hospitalisation, intensive care admissions and mortality.^{2,5,9,12-15} The techniques to improve airway clearance can reduce complications related to the accumulation of secretions and hospitalisation rates to a minimum.^{3,15,16} However, in patients with chest wall deformity, physiotherapy and manually assisted expectoration techniques can be ineffectual.¹⁷ These techniques are also time-consuming and require an appropriately trained carer.

Mechanical Insufflation-exsufflation (MIE) is a mechanically assisted cough technique. It gradually applies positive pressure to the airways, followed by a rapid change due to negative pressure. It can be applied using a face mask or a tracheotomy tube.^{2,5} In 1953, the first device for mechanical assistance of coughing was marketed. Since then, various studies have confirmed its efficacy. MIE is considered more effective than other reference techniques to increase the coughing mechanism.^{4,5,7} In spite of ever greater experience and published results on this technique, only limited data is available on its indications for home use, as also on compliance and safety.

The aim of this study is to describe the indications for home treatment using MIE, as also to determine its safety and compliance in patients with NMD.

Material and Methods

Since 1989, the Pneumology Service of the Sant João Hospital has an ambulatory clinic for NMD in collaboration with the Neurology Service. This clinic offers respiratory care to many patients suffering from NMD.

IEM devices have been used for home care since February 2005. The device was prescribed for continuous daily rent. These devices are included in the respiratory material offered to patients according to specific clinical indications. This material includes instruments for ventilation, monitoring, and control of secretions and is hired by a home care private company that also provides regular home visits by health professionals. The MIE device and a portable oximeter have a daily rental cost of 10 Euros. All the expenses caused by this home treatment are the responsibility of the Hospital. All the strategies related to clinical assessment of patients, adaptation of devices and programs for carer training are carried out by professionals from the Pneumology Service of the NMD ambulatory clinic.

All the patients prescribed home MIE were included in this study. An observational analysis was carried out with a 4 year follow-up. The patients were recruited in the Pneumology Service from those on the multidisciplinary NMD clinic. The protocol was approved by the Investigation Committee of the Hospital and the study was carried out according to the ethical directives for investigation in humans and the principles of the Helsinki Declaration (1975, revised in 1983).

Patients

Patients were studied from February 2005 to February 2009. Home MIE was prescribed for 21 NMD patients (15 men) of a median age of 58 years (27-72 years). The diagnosis included bulbar and non-bulbar amyotrophic lateral sclerosis (ALS), Duchenne muscular dystrophy (DMD) and other NMD. The demographic data of the population studied are described at table 1. Inclusion criteria were as follows: diagnosis of NMD, basal assisted < 270 l/min and continuous dependence on mechanical ventilation (24 hours a day). For the prescription of home MIE the decisive criteria was insufficient assisted PCF, after a technique of air entrapment (in patients with non-bulbar ALS) in combination with abdominal thrust. All patients with NMD receiving mechanical ventilation through a tracheotomy were also included.

Patients who could not cooperate or did not receive dedicated care were excluded. All patients depended on continuous home

Table 1
Characteristics and prior lung function in patients before applying mechanical insufflation-exsufflation protocol

Diagnosis	Amyotrophic lateral sclerosis	Duchenne muscular dystrophy	Other neuromuscular diseases	Multiple sclerosis	All
Patients, number	15	2	3	1	21
Age (onset of respiratory deterioration)	62 (46-72)	32 (30-34)	35 (27-36)	68	58 (27-72)
Duration of symptoms, months	26 (9-90)	83 (27-139)	38 (8-271)	7	29 (7-271)
FVC (l and predictable%)	0.99 (0.4-1.93) 32.5% (15-48%)	0.4 (0.26-0.54) 9.5% (7-12%)	0.6 (0.26-0.84) 14% (7-20%)	0.7 29%	0.81 (0.26-1.93) 27.5% (7-48%)
FEV ₁ (l and predictable%)	0.95 (0.3-1.76) 34% (7-56%)	0.37 (0.24-0.49) 10% (7-13%)	0.57 (0.24-0.84) 16.5% (7-23%)	0.67 33%	0.72 (0.24-1.76) 28% (7-56%)
MIP (cmH ₂ O)	31 (0-45)	14 (0-28)	16 (0-28)	ND	28 (0-45)
MEP (cmH ₂ O)	27 (0-56)	15 (3-27)	21 (3-27)	ND	22 (0-56)
Spontaneous PCF (l/min)	60 (0-250)	90 (80-100)	115 (80-150)	40	60 (0-250)
Assisted PCF (l/min)	160 (0-260)	170 (160-180)	192.5 (130-250)	80	160 (0-260)
NIV, No. of patients	10	2	2	1	15
Tracheotomy, No. of patients	5	0	1	0	6
Time undergoing continuous ventilation support, months.	24.5 (9-51)	81.5 (24-139)	37 (7-271)	6	29 (7-271)

FEV₁ indicates forced expiratory volume in first second; MEP, maximum expiratory pressure; MIP, maximum inspiratory pressure; NIV, non invasive ventilation; PCF, peak cough flow.

mechanical ventilation through a volume limited time-cycled ventilator (mean current volume 1,000 ml). At the beginning of the study, 6 patients had tracheotomy tubes and 15 patients used continuous VNI combining ventilation through a mouthpiece during the day and ventilation with a face-mask during the night.

Before entering the ambulatory MIE protocol, 20 patients had come to the emergency service due to respiratory complications related to the accumulation of secretions (median 3 hospitalisations per patient) and 12 patients had required hospitalisation due to respiratory infections (median 2 hospitalisations per patient).

Determinations

All patients underwent pulmonary function, respiratory muscle force and PCF screening. Pulmonary function was determined by spirometry (Vmax 229, Autobox, SensorMedics), registering the maximum expiratory volume during the first second of forced expiration (FEV₁) and forced vital capacity (FVC). All lung function tests were carried out in a sitting position according to reference procedures defined by the ATS-ERS 2005 work group.¹⁸

Maximum inspiratory pressure (MIP) and maximum expiratory pressure (MEP) were determined with a portable pressure manometer with an occludable mouthpiece (Micro RPM, Micromedical Limited) during maximum inspiratory and expiratory manoeuvres through the mouthpiece starting with the residual volume (MIP) and from total lung capacity (MEP). These determinations were performed according to reference procedures defined by the ATS-ERS 2002 work group.¹⁹ The manoeuvres were repeated until three determinations with a variability < 5% were obtained.

The PCF was only determined in non-tracheotomised patients by requesting the patient in a sitting position to carry out a cough manoeuvre beginning by total lung capacity (TLC) by means of a standard (Assess Healthscan products, Inc.) flow-meter connected to a face-mask.

Maximum insufflation capacity (MIC) was assessed by spirometry requesting the patient to retain 2 consecutive air volumes supplied by a manual resuscitator bag (air entrapment technique).²⁰ Bulbar deterioration and glottic dysfunction were evaluated in non-tracheotomised patients by assessing the MIC/FVC ratio. It was considered that patients with a ratio # < 1 presented bulbar dysfunction. For patients with appropriate bulbar function assisted PCF is determined after the air entrapment test with synchronised manual abdominal compression synchronised with the cough effort.

Since the patients with bulbar dysfunction could not carry out the air entrapment test, assisted PCF was only determined by application of manual abdominal compression.

Protocol for Home MIE

Home MIE was carried out using the Cough-Assist® (Philips, Respironics, Inc) device using an oronasal mask or a non-fenestrated tracheotomy tube with an inflated cuff. Both patients and carers were trained in the use of a portable pulse-oximeter (*Nonin 9500 oximeter Minneapolis Plymouth USA*) and given information on the use of MIE in their homes, in which a MIE session was applied in each episode of SpO₂ < 95%, until a value > 95% was obtained.²³ For better tolerance periods of rest with ventilation between sessions were applied. Before the prescription of home MIE, all the patients and carers in our service were convened to receive specific training on the technique with a specialized respiratory physiotherapist. Training included device management (adjustments and circuit connections) and a practical seminar with clinical simulations, and also detection of clinical signs necessary to determine efficacy. Each session consisted of 6-8 cycles of insufflation-exsufflation with mean pressures of 40 to -40 cmH₂O. The duration of each cycle was 3 seconds for insufflation, 2 seconds for exsufflation and 4 seconds for the post-exsufflation pause. During the exsufflation phase, patients were trained to cough, at the same time a carer applied abdominal compression. The adaptation of the technique was gradual, pressure was increased progressively to obtain an adequate chest expansion at a comfortable level to eliminate secretions.

Home MIE was always administered by non-professional trained carers (family members or private assistants) with the support of a health care professional with experience from a home care private company (nurse or respiratory physiotherapist). In case of doubt, the carer called the health professional to resolve the problem or to help carry out the technique in difficult situations. Both patients and carers were trained to detect early signs of respiratory failure or respiratory infections and were instructed to contact service staff (pneumologist or respiratory physiotherapist) on appearance of the first sign. Whenever dyspnoea increased, secretions accumulated or a value of SpO₂ < 95% persisted, in spite of continuous use of the ventilator and an aggressive home technique (with the support of expert health carers), the patients were instructed to come in to the emergency service (ES) of the local hospital. The aim of the treatment

Table 2
Compliance with mechanical insufflation-exsufflation

Diagnosis	Bulbar amyotrophic lateral sclerosis	Non-bulbar amyotrophic lateral sclerosis	Duchenne muscular dystrophy	Other myopathies	Multiple sclerosis	All
Daily users	7	2	0	1	0	10
Intermittent users	3	3	2	2	1	11

was defined as satisfactory control of acute dyspnoea or secretion accumulation, confirmed by pulse-oximetry data ($\text{SpO}_2 > 95\%$).

Variables Analysed

A descriptive statistical analysis was performed using the SPSS 14.0 database for Windows. Data presented are function on diagnosis, symptom duration, time on mechanical ventilation, spirometry, force of respiratory muscles and assisted and non-assisted PCF. Compliance, tolerance and efficacy were assessed based on daily frequency of MIE use, number of complications, number of visits to the ES due to episodes of secretion accumulation and number of hospitalisations related to airway infections. Clinical files were analysed to assess the number of visits to the ES and the number of hospitalizations prior to the MIE protocol.

Intolerance and adverse effects of the technique were also examined.

Results

A total of 21 patients with NMD undergoing continuous mechanical ventilation (6 with tracheotomies) and home MIE treatment were studied. The diagnoses were the following: Amyotrophic lateral sclerosis (ALS) ($n = 5$), Duchenne muscular dystrophy (DMD) ($n = 2$), other NMDs ($n = 3$) and multiple sclerosis (EM) ($n = 1$) (table 1). Other NMDs included a heterogeneous group of diseases: Myopathy due to cytoplasmic inclusion bodies, type 2 spinal muscular atrophy and non-classified myopathies. Lung function prior to MIE can be seen at table 1.

Median time of onset of respiratory deterioration of the patients were 29 months (7-271 months) and had been using home ventilation support treatment for a median of 29 months (7-271 months) before their inclusion in the protocol (table 1). As to the ALS patients, at the beginning of the program, there were 6 patients with severe bulbar dysfunction. During follow-up, there was disease progression, so that, at the end of treatment, 10 patients presented severe bulbar dysfunction, 5 of them had undergone tracheotomy (5 rejected it). There was also a patient with non-classified myopathy, ventilated through a tracheotomy, without bulbar dysfunction, that rejected decannulation to VNI.

Compliance with MIE is shown in table 2. This technique was used daily by 10 patients (7 with bulbar ALS and 1 with non-classified myopathy) (table 2). The 6 patients with tracheotomy (5 with bulbar ALS and 1 with non-classified myopathy) and 4 patients undergoing VNI (2 with bulbar ALS that rejected tracheotomy and 2 with non-bulbar ALS) used the technique daily. The data related to daily frequency of sessions in the groups of daily users are shown in table 3. In this group, the patients with tracheotomies used MIE more times a day than those with VNI (table 3).

The technique was used intermittently by 11 patients (table 2). These patients only used the device during acute exacerbations, such as infection of the airways or during individual episodes of secretion accumulation. When the exacerbation was resolved of the secretions reduced to the point of maintaining a $\text{SpO}_2 > 95\%$, its use was interrupted.

In general, the carers considered the device was effective.

In 8 patients the early application of MIE (guided by oximetry data) prevented visits to the ES due to an accumulation of secretions

Table 3
Daily users of mechanical insufflation-exsufflation

Patients with ventilator support	Median (min-max)	No. of patients
Users of ventilations that underwent tracheotomy	6 (3-6) times a day	6 (100%)
Users of non-invasive ventilation	3 (1-4) times a day	4 (26.7%)
All	4.5 (1-6) times a day	10 (47.6%)

and the device inverted the episode of desaturations and normalized SpO_2 without the need to administer supplementary oxygen. The 8 patients and their respective carers reported that, if they had not had access to the device, they would have had to come in to the emergency service.

In 4 patients (3 bulbar ALS and 1 non-bulbar ALS), home MIE was not sufficient to normalise oxygen saturation and required hospitalisations to treat accumulated secretions during acute respiratory airway infections (table 4). Of the 3 bulbar ALS patients, one of them had undergone a tracheotomy and the other 2 had constantly rejected it.

Home MIE was well tolerated and there were no complications related to treatment (table 4).

During the study period, 4 patients with bulbar ALS died due to disease progression (table 4).

Discussion

The efficacy of MIE to increase PCF and improve the efficacy of cough manoeuvre has already been demonstrated.^{1-4,7,13,14,21,22} Chatwin et al compared different interventions (cough assisted by physiotherapy, cough assisted by VNI, cough assisted by exsufflation and MIE) with non-assisted cough and showed that MIE was the technique associated with the greatest increase in PCF.⁴ Oximetry data has shown the usefulness of this technique since it makes it possible to detect a sudden decrease of oxygen saturation as a consequence of a mucous plug.^{2,3,15,21} The patients described in this study suffered from severe ventilation failure and continuously depended on mechanical ventilation (24 hours a day). They had all used volume limited time-cycled ventilation for many years before inclusion in the study.

This study was based on the protocol proposed by Bach et al, that consists in home treatment with a VNI support, oximetry monitoring during 24 hours and the use of MIE guided by the data from this ($\text{SpO}_2 < 95\%$).^{3,7,15,23,24}

In this study home MIE was carried out by non-professional carers of the patients with the support of health professionals trained in home care.

Few studies have described home respiratory treatment with MIE in patients with NMD, although many studies describe the efficacy of the combination of cough assistance techniques, manual and mechanical, in acute in-hospital situations.^{8,9,21} Although MIE carried out by non-professional carers has been considered effective and well tolerated, it causes certain controversy and we still require more data on its home use in the long term.^{15,21,23,24}

In our strategy, the main condition for effective home care after hospital discharge was the presence of appropriately trained and

Table 4
Follow-up and results of mechanical insufflations-exsufflation

Diagnosis	Bulbar amyotrophic lateral sclerosis	Non-bulbar amyotrophic lateral sclerosis	Duchenne muscular dystrophy	Other myopathies	Multiple sclerosis	All
Patients, No.	10	5	2	3	1	21
Time undergoing MIE, months	6.5 (3-41)	19 (12-38)	7.5 (3-12)	16 (6-16)	4	12 (3-41)
Intolerance MIE	0	0	0	0	0	0
Complications due to MIE	0	0	0	0	0	0
Visits to the emergency service (No. of patients/No. of events)	3/5	1/1	0/0	0/0	0/0	4/6
Hospitalisations (No. of patients/No. of events)	3/3	1/1	0/0	0/0	0/0	4/4
Deaths, No. of patients	4	0	0	0	0	4

MIE indicates mechanical Insufflation-exsufflation.

motivated carers. This study supports the importance of an appropriate early training phase, administered to both patients and carers in the hospital, as has also been proposed by Tzeng et al.¹⁵ Furthermore, This study confirmed that carers with appropriate training and motivation can detect respiratory worsening and effectively use a home MIE protocol. Patients with neuromuscular diseases, especially ALS, can suffer progressive disease with early decrease of respiratory muscle force and cough deterioration, both associated with premature death.^{3,9,14,25,26} The natural course of NMD does not make it possible to clarify the influence of home MIE on visits to the ES/hospitalizations. The number of episodes before and after the home MIE protocol is not comparable. Indeed, before beginning home treatment, the patients' cough reflex was more powerful and therefore, they suffered fewer episodes of secretion accumulation. In contrast to other NMD, ALS also includes glottic muscle control. In general, in patients with non-bulbar NMD non-invasive support treatment may be used and they have a higher survival rate.^{3,5,16} However, the almost inevitable progression to bulbar dysfunction is one of the more negative characteristics of this disease and the main reason due to which, in contrast to other NMDs, tracheotomy becomes necessary to prolong survival.^{1,3,6-9,12,13,16,23,24,26,27} The MIC/FVC ratio has been widely used to assess bulbar function, due to the fact that appropriate glottic function is necessary to accumulate consecutive volumes of air to achieve MIC.²⁰ Furthermore, a severe bulbar dysfunction is also responsible for dysfunction of the higher respiratory airway muscles in this group of patients.^{2,4,13,23,25} Sancho et al have identified 2 types of bulbar ALS patients: those that only suffer from failure of glottis closure that cannot entrap air but in which MIE can be effective and those that present a dynamic collapse of the upper respiratory airways in which MIE is not effective and can even cause risk.²⁵ In this study, ALS, the most usual diagnosis, is seen in a heterogeneous group: from a non-bulbar disease with inadequate PCF to a severe bulbar disease that requires tracheotomy. This in itself represents an additional problem concerning patient acceptance or associated problems (local inflammation, increase of secretions and infections).^{2,3,12,15,26,28} Indeed, although it was offered to all patients with severe bulbar dysfunction, some continued to reject it and preferred to continue with VNI and the MIE protocol. Bach et al have also described the fact that patients with severe bulbar ALS can receive support treatment with the combined use of VNI and MIE, which delays tracheotomy, as long as they can maintain an oxygen saturation > 95% with ambient air.²³ However, these patients required careful regular supervision to anticipate the failure of a non-invasive strategy, so that therapeutic options can be examined and analysed with patients and their families, so that decisions can be made beforehand and not at the time of a respiratory crisis. Farrero et al have suggested that a follow-up every 3 months or on patient demand makes it possible to opportunely recognise disease progression.¹⁶

In this study, each MIE session consisted of 6-8 cycles of insufflations-exsufflations with mean pressures of 40 to -40 cm H₂O, titrated according to patient tolerance. Chatwin and Sivasothy have considered that lower pressures are more comfortable and involve fewer risks.^{4,17} However, the adjustments used in this study are widely preferred due to their effects on patient welfare and their efficacy and are also suggested by the manufacturers.^{1,3,8,24,29} In general, they are well tolerated and have been considered the most effective means of obtaining higher values of PCF with practically no complications.^{1,3,5,7,8,21,24} In spite of this, we paid special attention to careful individual titration of the MIE pressure, to obtain maximum chest expansion, respecting welfare, which can justify the tolerance and absence of complications seen in this study. The application of MIE was adjusted to the needs of each patient in as far as frequency of use, considering daily and intermittent users. With intermittent users it was not possible to register the number of MIE sessions per month or week because compliance was not regular and depended on the number and severity of respiratory exacerbations. In contrast, daily users reported that they used the device every day, independently of exacerbations, both for secretion control and for lung insufflation and to revert episodes of SpO₂ < 95%. This study also shows that patients that had undergone tracheotomy used MIE daily and more times a day than patients with VNI. This fact may be due to local inflammation and the increase of secretions related to tracheotomy. We also found that some patients with bulbar ALS, incapable of air entrapment, did not only use MIE as a technique for cough assistance, but also for lung insufflation.

According to this study, both patients and carers described greater efficacy in clearance of airway secretions with this home protocol. Its early application, guided by oximetry data with normalisation of oxygen saturation with ambient air, prevented visits to the Emergency Service due to secretion accumulation. Patients stated that, during these episodes, without MIE, they would have had to go to hospital due to their acute respiratory problems. Furthermore, there were very few episodes in which home MIE was not sufficient to resolve the problem of secretion accumulation and made it unnecessary to visit the ES or to require hospitalisation. None of these patients required intubation. Indeed, it would seem that the protocol reduces the risk of airway infection and prevents both visits to the ES and hospitalizations. Bach et al have reported that patients with NMD dependent on VNI that used MIE guided by oximetry data can be treated at home without risk or need of hospitalisation.^{3,6,7,13,14,15}

This study did not show any complications related to use of the device. Potential complications are very infrequent and include abdominal distension, increase of gastroesophageal reflux, haemoptysis, chest and abdominal discomfort, acute cardiovascular events, barotraumas and pneumothorax.⁵ Bach et al have not described any complications in more than 500 batches of MIE.⁷ Some simple but prudent measures to prevent complications are: brief

pauses between applications (to avoid hyperventilation), avoiding applications after meals, appropriate treatment of gastroesophageal reflux and reduction of insufflation pressure according to tolerance.⁵

During the study period, 4 patients with bulbar ALS died. The main cause of death was rapid progressive disease with severe bulbar dysfunction. These 4 patients had constantly rejected tracheotomy.

The main limitations of this study are the fact that it is observational, the reduced number of patients including the absence of a control group. However, shortly after beginning this protocol, the benefits of the treatment as far as efficacy in management of accumulated secretions, preventing visits to the Emergency Service and improvement of quality of life in these patients became evident. As a result, we consider that it would not be ethical to deprive the patients of a treatment that has been shown to be effective.

Conclusion

The most important conclusion of this study is that it is possible to treat patients with severe NMD with sufficient clearance of the secretions of their respiratory airways with home MIE based on careful training of non-professional carers. This is valid for patients with VNI and those with tracheotomies. According to the patients, greater use of MIE during respiratory infections can prevent visiting the emergency service.

This technique can be considered a useful complement to ventilation support in these patients.

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