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Yazışma Adresi
Correspondence Address

Abdurrahman Erdem BAŞARAN
Department of Pediatrics,
Division of Pulmonology,
Akdeniz University Faculty of Medicine,
Antalya, Turkey

erdembasaran15@hotmail.com

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Abdurrahman Erdem BAŞARAN
Department of Pediatrics,
Division of Pulmonology,
Akdeniz University Faculty of Medicine,
Antalya, Turkey
ORCID: 0000-0002-9092-6936

Ayşen BAŞARAN
Department of Pediatrics,
Division of Pulmonology,
Akdeniz University Faculty of Medicine,
Antalya, Turkey
ORCID: 0000-0001-7384-021X

Tuğba KAZLI
Department of Pediatrics,
Division of Pulmonology,
Akdeniz University Faculty of Medicine,
Antalya, Turkey
ORCID: 0000-0002-5412-782X

Suzan YILMAZ DURMUŞ
Department of Pediatrics
Division of Pulmonology
Akdeniz University Faculty of Medicine
Antalya, Turkey
ORCID: 0000-0001-7285-8928

Özgür DUMAN
Department of Pediatrics
Division of Neurology
Akdeniz University Faculty of Medicine
Antalya, Turkey
ORCID: 0000-0002-3313-8052

Şenay HASPOLAT
Department of Pediatrics
Division of Neurology
Akdeniz University Faculty of Medicine
Antalya, Turkey
ORCID: 0000-0003-3596-1816

Ayşen BİNGÖL
Department of Pediatrics
Division of Pulmonology,
Akdeniz University Faculty of Medicine
Antalya, Turkey
ORCID: 0000-0002-0886-3332

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The Effect of Mechanical Insufflation-Exsufflation Device on the Quality of Life and Hospitalization in Children with Neuromuscular Disorders

Mekanik İnsuflasyon-Eksuflasyon Cihazının Nöromusküler Hastalıkları Olan Çocuklarda Yaşam Kalitesine ve Hastane Yatışına Etkisi

ABSTRACT

Objective:

Respiratory complications are a significant cause of morbidity and mortality in patients with neuromuscular diseases (NMD). The aim of this study was to examine the demographic data of subjects with NMD using a mechanical insufflation-exsufflation (MI-E) device and the effect of an MI-E device on quality of life (QoL) and hospitalization.

Material and Methods:

The study included patients under 18 years with NMD followed up in the Pediatric Pulmonology Section between December 2019 and December 2020. Twenty-seven patients with NMD using an MI-E for at least 3 months were enrolled in the study group and 30 patients with NMD using only manuel airway clearance techniques were enrolled in the control group. Data were collected with respect to clinical and demographic characteristics, the MI-E device settings, ease of expectoration visual analogue scores (EE), and the QoL scores. Thirteen subjects, who used an MI-E device for more than 12 months, were assessed pre-and post-MI-E use hospitalization data for respiratory reasons.

Results:

The mean EE scores were significantly higher after MI-E device use compared to before. Among participants who were evaluated pre- and post-MI-E use there was a significant decrease in the number of hospital admissions and length of stay. The mean QoL scores of subjects diagnosed with spinal muscular atrophy (SMA) with tracheostomy in study group were determined to be significantly higher than SMA with tracheostomy in control group.

Conclusions:

The use of MI-E device showed a positive effect to EE, number of hospital admissions and length of stay in patients with NMD. In addition, it contributed positively of QoL scores in patients with SMA.

Key Words:
Mechanical insufflation-exsufflation, Neuromuscular disorder, MI-E device, Quality of life

ÖZ

Amaç:
Nöromusküler hastalığı (NMH) olan olgularda en önemli morbidite ve mortalite nedeni solunumsal komplikasyonlardır. Çalışmanın amacı Mekanik İnsuflasyon-Eksuflasyon (Mİ-E) cihazı kullanan NMH'li olguların demografik verilerini ve Mİ-E cihazının yaşam kalitesine ve hastane yatışına olan etkisini incelemekti.

Gereç ve Yöntemler:
Aralık 2019 ve Aralık 2020 tarihleri arasında Çocuk Göğüs Hastalıkları Polikliniği'nde NMH tanısıyla takip edilen 18 yaşından küçük hastalar çalışmaya dahil edildi. En az 3 aydır Mİ-E cihazı kullanan 27 NMH'li hasta çalışma grubuna ve sadece manuel solunum yolu temizleme teknikleri kullanan 30 hasta kontrol grubuna alındı. Klinik ve demografik özellikleri, Mİ-E cihazı ayarları, balgam çıkarmadaki kolaylık vizüel analog skorları (BÇK) ve yaşam kalitesi skorlarıyla ilgili veriler toplandı. Mİ-E cihazını 12 aydan uzun süre kullanan 13 olgunun Mİ-E cihazı öncesi ve sonrası solunumsal nedenlerle hastaneye yatış verileri değerlendirildi.

Bulgular:
Mİ-E cihazı kullanımı sonrası BÇK skorları cihaz kullanımı öncesine göre istatiksel anlamlı yüksekti. Mİ-E cihazı kullanımı öncesi ve sonrası değerlendirilen olgularda hastaneye yatış sayısında ve süresinde istatistiksel anlamlı azalma vardı. Çalışma grubu içerisindeki trakeostomili spinal musküler atrofi (SMA) tanılı olguların yaşam kalitesi skorları ortalaması kontrol grubundaki trakeostomili SMA'lı olgulara göre istatistiksel anlamlı yüksek saptandı.

Sonuç:
Mİ-E cihazı kullanımı NMH'li hastalarda BÇK'ya, hastaneye yatış sayı ve süresine olumlu katkı sağladı ve SMA'lı hastalarda yaşam kalitesine olumlu katkıda bulundu.

Anahtar Sözcükler:
Mekanik İnsuflasyon-Eksuflasyon, Nöromusküler hastalık, Mİ-E cihazı, Yaşam kalitesi

INTRODUCTION
Respiratory complications are a significant cause of morbidity and mortality in patients with neuromuscular diseases (NMD) (1). The inability for full expansion of the chest wall due to weakness of the respiratory muscles leads, over time, to chest wall stiffness, chronic hypoventilation, atelectasis, and insufficient cough (2). An accompanying weak or ineffective cough causes recurrent respiratory tract infections (RTIs), eventually resulting in respiratory failure.
When the intercostal and abdominal muscles are not able to produce sufficient thoraco-abdominal pressure, patients with NMD should be supported with manual and/or mechanical methods for coughing (3-5). Manuel methods include active cycle of breathing technique, postural drainage, or chest physical therapy and mechanical methods include mechanical insufflation-exsufflation (MI-E), intrapulmonary percussive ventilation, and high-frequency chest wall compression (6). MI-E devices increase peak cough flow by increasing expiratory air flow with simultaneous cough. Increased peak cough flow provides clearance from the mouth or tracheostomy through central airway movement of secretions. A report from two new international panels, which included specialists on this subject, reported that MI-E devices are an expensive but ideal cough support technique for individuals severely affected by neuromuscular diseases; however, the management approach also depends on availability of equipment and local expertise, which may vary substantially at a global level (7, 8).
It is inevitable that there will be physical, social, and emotional effects on children with NMD with restricted movement and the feeling of respiratory muscle weakness. Although several studies found that quality of life (QoL) is impaired in spinal muscular atrophy (SMA) and Duchenne muscular dystrophy (DMD) patients, especially in the physical domain, Morrow et al. stated in a recent meta-analysis that we need additional studies to measure the effect of cough augmentation techniques on morbidity, mortality, and QoL for people with NMD (9). To our knowledge, no previous studies have examined yet the effect of the MI-E device on the QoL of NMD patients using validated QoL tools in the literature. Hovewer, there are only two studies that looked at the effect of MI-E on hospitalization rates in pediatric patients with NMD (10, 11).
The current study mainly aimed to examine the demographic data of subjects with NMD using an MI-E device and retrospectively analyze the number of admissions to hospital and the Intensive Care Unit (ICU) for respiratory reasons before and after initiation of MI-E. Second, we aimed to prospectively evaluate the effect of an MI-E device on QoL by using a validated questionnaire.

MATERIALS and METHODS

Subjects
This was a prospective cross-sectional designed study. We enrolled patients with NMD under 18 years followed up in the Pediatric Pulmonology Section of Akdeniz University Hospital between December 2019 and December 2020. The study group consisted of 27 patients with NMD (17 SMA and 10 DMD) who had been using the MI-E device (Philips Respironics, Philip Healthcare; Best, The Netherlands) for at least three months. In order to evaluate the effect of the MI-E device on the quality of life, 30 patients with NMD (20 SMA and 10 DMD) who used only manuel airway clearance techniques with similar age, gender, and respiratory parameters served as the control group.
We reviewed the medical records to obtain demographic characteristics, diagnosis, age at diagnosis, the presence of tracheostomy, non-invasive/invasive respiratory support, data on hospital presentations, and number of days spent in hospital and the intensive care unit for respiratory reasons. Also, information about the duration of MI-E use, insufflation-exsufflation pressures, inspirium-expirium-pause durations, how many

times per day and days per week the MI-E is used in the presence or absence of RTIs, any side-effects seen associated with use, ease of expectoration visual analogue scores (EE) which asked 'how easy was it to cough and expectorate sputum?' before and after starting to use an MI-E device (0-10 points; 0=extremely difficult, 10= extremely easy) and whether or not any adjustments had been made to the MI-E device settings were collected from the parents of the study group using an online questionnaire created through Google forms (12).

According to our protocol, for subjects with audible weak cough, retained secretions secondary to respiratory muscle weakness, difficulty clearing secretions, history of pneumonia, prolonged or frequent RTIs, or reduced respiratory function tests, we begin manuel airway clearance techniques at home. When efficacy or technique (e.g., due to young age) is insufficient, we switch to MI-E device if the family can obtain the device, whether by paying the price of the device itself or through a charity campaign. Device settings (insufflation/exsufflation pressure beginning with +14 cm H2O to -14 cm H2O and maximum pressure of +55 cm H2O to -55 cm H2O, insufflation/exsufflation/pause time were each 0.8–3 seconds (sec)) are customized according to tolerance, amount of secretions, and chest auscultation. We teach caregivers to perform three sessions, each with five cough cycles, at a frequency of at least twice a day and more frequently during RTIs.

This research complies with all the relevant national regulations, institutional policies and is in accordance with the tenets of the Helsinki Declaration, and has been approved by the Akdeniz University Ethical Committee, (Approval no KAEK-29; date 13/1/2021).

QoL measure

The Peds QL Neuromuscular Module, version 3 questionnaire was completed online by the parents and by the children who were capable of completing the questionnaire independently. This questionnaire has been validated for children with neuromuscular disease and consists of 25 items; 17 items with an emphasis on physical functioning, 3 items related to communication, and 5 items related to family resources. For each item, the parent/child is asked to rate the impact of a particular problem over the past month. Each multiple choice answer is scored from 0 (never a problem) to 4 (almost always a problem). Item scores (Quality of Life scale-total points (QoL-TP), Quality of Life scale–Neuromuscular disease total points (QoL-NTP), Quality of Life scale–communication total points (QoL-CTP) and Quality of Life scale– family resources total points (QoL-FRTP)) are reversed and converted linearly to a scale from 0 to 100 (0 = 100, 1 = 75, 2 = 50, 3 = 25 and 4 = 0), with higher scores indicating a higher QoL (13, 14).

In order to evaluate the effect of the MI-E device on QoL, the Peds QL questionnaire results of 17 SMA patients in the study group and 20 SMA patients in the control group who had no significant differences in terms of SMA type, age, gender and tracheostomy status were compared. Additionally, the Peds QL questionnaire results of 10 patients with DMD in the study group and 10 patients with DMD in the control group who were matched for age, gender, and respiratory parameters were compared.

Hospitalization data

In order to eliminate the effect of group variables on the results, the pre- and post-MI-E use hospitalization data on presence, number, and duration of hospitalizations and intensive care unit admissions of the subjects for respiratory reasons who used an MI-E device for more than 12 months were evaluated.

Statistical Analysis

Data obtained in the study were analyzed using SPSS vn. 23.0 software. Descriptive statistics are presented as frequency, percentage, mean, standard deviation, and minimum and maximum values. Conformity of the data to normal distribution was assessed with the Shapiro-Wilk test. In the analysis of differences between the two groups, the Independent Samples t-test was used if the data fit the normal distribution and the Mann-Whitney U Test was used if the data were not normally distributed. The Wilcoxon signed rank test was applied to compare the differences before and after MI-E device use with respect to EE scores, and the number and duration of admissions to hospital and ICU. To evaluate relationships between the variables, Pearson correlation analysis was applied. A value of $p<0.05$ was accepted as statistically significant.

RESULTS

There were 27 patients in the study group; 16 (59.3%) males and 11 (40.7%) females with a mean age of 9.39±6.76 years. They were diagnosed with SMA type 1 in 15 subjects, SMA type 2 in 2 subjects, and DMD in 10 subjects. There were 30 patients in the control group; 22 (73.3%) males and 8 (26.7%) females with a mean age of 8.05±6.18 years. They were diagnosed with SMA type 1 in 17 subjects, SMA type 2 in 3 subjects, and DMD in 10 subjects. No significant differences were determined between SMA patients in the study and control groups in terms of SMA type, age, gender, and tracheostomy status (Table I).

Table I: Demographic and clinical features of patients with SMA in the study and control groups.

	SMA subjects in study group (n=17)	SMA subjects in control group (n=20)	p
SMA type Type 1, n (%) Type 2, n (%)	 15 (88.2) 2 (11.8)	 17 (85) 3 (15)	0.77
Age (years) *	5.14±4.60	4.17±2.72	0.45
Gender, male (%)	6 (35.3)	13 (65)	0.10
Age at diagnosis* (months)	5.85±3.51	9.12±8.78	0.13
Presence of tracheostomy, n (%)	13 (76.47)	11 (55)	0.30
Requirement for intermittent oxygen support, n (%)	5 (29.4)	9 (45)	0.49
Non-invasive support, n (%)	2 (11.7)	2 (10)	0.99

SMA: Spinal muscular atrophy, *Values presented as median±SD.

Additionally; there were no significant differences between DMD patients in both groups in terms of age and respiratory parameters (Table II).

Table II: Comparison of demographic and clinical characteristics and respiratory parameters of DMD cases in the study and control groups.

	DMD subjects in study group (n=10)	DMD subjects in control group (n=10)	p
Age (years) *	16.6±1.57	15.8±2.69	0.43
Age at diagnosis* (months)	57.6±16.7	40.2±26.3	0.09
FVC (% predicted)*	50.4±29.0	60.1±22.9	0.48
FEV1 (% predicted)*	57.1±30.3	67.1±21.3	0.47
Cough Peak Flow (L/min)*	195.4±63.4	208.5±54.4	0.67
Ambulatory status, required assistance, n(%)	9 (90)	8(80)	0.50
Non-invasive support, n (%)	3 (30)	1 (10)	0.58

DMD: Duchenne muscular dystrophy, FVC: Forced vital capacity, FEV1: First second of forced expiration. *Values presented as median±SD

The mean EE score of the 27 subjects using MI-E device was determined to be significantly higher after MI-E device use compared to before (9.11±1.09 vs 4.37±3.40, p<.001).

MI-E device use data

The mean duration of use of the MI-E device was 12.04±9.52 (3-36) months. The mean age at which they started using MI-E was 100.6±80.3 months in all subjects, 49.0±51.6 months in SMA subjects, 188.4±17.7 months in DMD subjects.

The mean insufflation pressure used was 21.19±8.95 (14-50) cmH2O, exsufflation pressure was 24.63±12.05 (14-55) cmH2O, insufflation time was 1.49±0.48 (0.8-2.5) sec, exsufflation time was 1.61±0.60 (0.8-3) sec, and pause duration was 1.6±0.6 (1-3) sec. The MI-E device was used for a mean of 5.33±2.27 days per week and 2.26 ± 1.46 (0-7) times per day when the subject was not ill, and for a mean of 6.44±1.37 days per week and 3.67 ± 1.54 (1-7) times per day during periods of respiratory infection. The MI-E device was used via the tracheostomy by 13 (48.1%) subjects and fixed to the face mask in 14 (51.9%) subjects. The device mode used was basic automatic in 17 (63%) subjects, manual in 6 (22.2%) subjects, triggered auto in 2 (7.4%) subjects, and timed auto in 2 (7.4%) subjects. Changes were made to the MI-E device settings, including during periods of use in 13 (48.1%) subjects, and no changes were made in 14 (51.9%) subjects. No complications associated with MI-E device use were determined in any of the subjects.

Hospitalization data

Thirteen cases in the study group were using the MI-E device for more than 12 months. Of these cases, 6 had SMA type 1, 1 had SMA type 2, and 6 had DMD. Eight (61.5%) of the cases were male and 5 (38.5%) were female, with a mean age of 11.2±6.8 years. In the year after using the MI-E device, there was a significant decrease in the number of hospital admissions (p=.01) and length of stay (p=.01) compared to the year prior to using the MI-E device. No significant differences among groups were determined with respect to hospital admission within the last year (p=.063), admission to ICU (p=.37), number of ICU admissions (p=.15), and length of stay in ICU (p=.08) (table III).

Table III: Hospitalization before and after the implementation of MI-E device.

	Before MI-E use (n=13)	After MI-E use (n=13)	p
Admission to hospital in the last 1 year	69.2%	30.8%	0.06
Number of hospital admissions in the last 1 year*	1.61±0.51	0.30±0.13	**0.01**
Length of stay in hospital in the last 1 year (days)*	19.84±6.10	1.84±0.83	**0.01**
Admission to ICU in the last 1 year	30.8%	7.7%	0.37
Number of ICU admissions in the last 1 year *	0.61±0.38	0.07±0.27	0.15
Length of stay in ICU in the last 1 year (days) *	12.15±5.88	0.15± 0.55	0.08

ICU: Intensive care unit, MI-E: Mechanical insufflation-exsufflation. *Values presented as mean±S.D (Table III).

Proxy-reported QoL Scale points

When the QoL scores of all SMA subjects in the study and control groups were evaluated, there was a significant difference between the two groups with respect to QoL-NTP (56.16 ± 17.61 vs 42.57±24.93, p=.03), and no significant differences with respect to QoL-TP (p=.191), QoL-CTP (p=.80), and QoL-FRTP (p=.34).

When the QoL scores of SMA subjects with tracheostomy in the study (Five (45.4%) of the cases were male and 6 (54.6 %) were female, with a mean age of 3.2±1.5 years) and control groups (Seven (63.6%) of the cases were male and 4 (36.4 %) were female, with a mean age of 3.4±1.9 years) were evaluated, the mean QoL-TP (50.73±18.28 vs 29.27±20.89, p=.016) and QoL-NTP (57.06±18.95 vs 31.14±19.41, p=.002) were significantly higher in the study group than in the control group.

No significant differences were determined between the proxy-reported QoL Scale points of the DMD subjects in the study and control groups with respect to the QoL-TP (p=.17), QoL-NTP (p=.12), QoL-CTP (p=.15), and QoL-FRTP (p=.59).

Table IV: Average scores of proxy-reported PedsQL 3.0 NMM across groups.

Variable	SMA (n=15) in study group	SMA (n=20) in control group	p	SMA+ tracheostomy in study group (n=11)	SMA+ tracheostomy in control group (n=11)	p	DMD (n=10) in study group	DMD (n=10) in control group	p
QoL-TP*	49.9±18.1	41.0±25.6	0.19	**50.7±18.2**	**29.3±20.9**	**0.016**	47.3±14.9	56.7±15.0	0.17
QoL-NTP*	**56.1±17.6**	**42.5±24.9**	**0.03**	**57.0±18.9**	**31.1±19.4**	**0.002**	43.8±17.0	57.0±19.5	0.12
QoL-CTP*	48.8±38.9	53.3±43.5	0.80	56.1±38.5	38.6±44.4	0.30	50.8±32.2	69.9±25.2	0.15
QoL-FRTP*	35.3±25.1	28.0±27.7	0.34	35.9±23.4	15.9±19.6	0.056	49.5±18.7	45.0±18.4	0.59

*Values presented as mean±S.D
Quality of Life scale- total points (QoL -TP), Quality of Life scale –Neuromuscular disease total points (QoL-NTP), Quality of Life scale – communication total points (QoL-CTP), Quality of Life scale – family resources total points (QoL-FRTP)
SMA: Spinal muscular atrophy; DMD: Duchenne muscular dystrophy

Self-administered QoL scale points

No significant differences were determined between DMD patients in the study and control groups with respect to the QoL-TP (p=.81), QoL-NTP (p=.95), QoL-CTP (p=.23), and QoL-FRTP (p=.70).

Correlations

A moderate negative correlation was determined between the number of days per week of MI-E device use when ill and the EE score before the use of the MI-E device (r=-.399, p=.03).
A moderate positive correlation was determined between the EE score after MI-E device use and the number of times per day of MI-E device use when not ill (r =.516, p=.006) and the number of days per week of MI-E device use when ill (r=.389, p=.04).
A moderate negative correlation was determined between the number of days per week of MI-E device use when ill and the length of stay in hospital (r=-.565, p=.04).

DISCUSSION

The results of this study demonstrate that the use of an MI-E device by subjects with NMD reduced the number and duration of hospitalizations, and in subjects diagnosed with SMA, especially those with tracheostomy, it provided a positive contribution to QoL. To the best of our knowledge, this is the first study to show the effect of MI-E device use by pediatric SMA subjects using validated QoL tools.
To date, there are two studies in the literature that have examined the effect of MI-E device use on hospital admissions in pediatric patients with NMD. Veldhoen et al. reported that the use of an MI-E device by children with NMD reduced hospital admissions and length of hospital stay associated with RTIs (10). In a study by Moran et al., although no reduction was determined in the number of presentations to the hospital by pediatric patients with NMD after starting to use an MI-E device, the length of stay in hospital was reduced in the 6th and 12th months of MI-E device use (11). In the current study, a significant reduction was determined in both the number (p=.01) and duration (p=.01) of hospitalizations with the use of the MI-E device by pediatric subjects with NMD. Although there were decreases in hospitalizations, admissions to ICU, and ICU stay duration, within the prior year after initiating use of an MI-E device, it was not statistically significant. However, a moderate negative correlation was determined between the number of days per week of MI-E device use when the subjects were ill and the length of hospital stay after starting to use the MI-E device. This finding was considered to support the increase in weekly use of the device while ill.
To the best of our knowledge, there is no previous study in literature that has examined the effect of the MI-E device on the QoL of NMD patients using validated QoL tools. However, Moran et al. reported that by decreasing the need for hospitalization, the use of the MI-E device made a positive contribution by improving the lifestyle of both the children and their parents (11). In the current study, the mean QoL-NTP of the children with SMA who used the MI-E device was significantly higher than that of the subjects who did not use the device. In addition, the mean QoL-TP and the mean QoL-NTP were significantly higher in the SMA subjects with tracheostomy using the MI-E device compared to those not using the device. This difference was not seen between subjects with DMD in study and control group. A significant increase was determined in the EE scores after the use of the MI-E device compared to before. This positive effect of the MI-E device on QoL was thought to be associated with the prevention of recurrent atelectasis by supporting ease of expectoration, the decreased need for hospitalization, and the positive effect on chest wall complications through the contribution of positive insufflation pressures (15). The reason for the determination of this difference, especially in SMA subjects with tracheostomy but not in DMD subjects, was thought to be due to the greater involvement of respiratory muscles in patients diagnosed with SMA, and, because of the younger age of the SMA patients, there was less co-operation and thus an insufficient degree of benefit was gained from other chest physiotherapy techniques (e.g., air-stacking using glossopharyngeal breathing). Also, as patients with tracheostomies are arguably weaker and more prone to respiratory failure, the MI-E tends to be more effective when administered via a tracheostomy, so they also might get more benefit from the MI-E device. In the literature, MI-E treatment is accepted as a significant aid to long-term mechanical ventilation (LTMV) (16, 17). In a study by Hov et al., which included pediatric patients with NMD or disease of central nervous system origin, it was reported that 56% of the patients using MI-E were receiving ventilator support at the same time (18). The combined use of MI-E and ventilator support was reported at the highest rate (83%) in SMA patients. Chatwin and Simonds reported that 96% of

patients were using both MI-E and LTMV (15). Consistent with these findings in the literature, of the 27 subjects using MI-E device in the current study, 18 (66.6%) were also receiving LTMV (5 non-invasive, 13 invasive).

The MI-E device has been used in NMD since 1954 for adults and children (19). Assisted coughing is a critical element of respiratory care for patients with SMA and may be the only way for them to cough and clear secretions (20). This device can completely remove bronchial secretions in six minutes (21). Its use has been proven positive especially in DMD patients with scoliosis. It has also been reported that MI-E devices could prevent hospitalizations and tracheostomy (22). In literature, the use of MI-E devices is recommended in NMD patients with cough peak flow (CPF) <160L/min, but there is no evidence or consensus related to the ideal device settings yet (7, 16). Hov et al. evaluated MI-E device settings in a study of 240 patients from 10 centres in 7 European countries and reported that lower MI-E pressures were used in younger children (23). There was also reported to be a wide range of pressures used; insufflation pressure minimum in the range of +10 cmH20 to +50 cmH20 and exsufflation pressure between -10 cmH20 and -60 cmH20. In the same study, half of the children used asymmetric pressures, and in the majority of these the exsufflation pressure was higher than the insufflation pressure. Just as there are reports in the literature advocating the use of high MI-E device pressures there are also studies showing the benefits of low pressures such as 15 cmH20 (24-28). In the current study, the mean insufflation pressure was found to be 21 cmH20, mean exsufflation pressure 24 cmH2O, mean insufflation time 1.49 sec, mean exsufflation time 1.61 sec, and mean pause time 1.6 sec. These values were consistent with the findings in the literature showing that asymmetric pressures were used more frequently and exsufflation pressures were higher than insufflation pressures. Almost half of the subjects in this study had made changes to the device settings while using it and no complications were determined in any subject. With these values, there was a positive contribution to the number and duration of hospitalizations and to EE scores, and considering that MI-E devices may very rarely cause pneumothorax it can be recommended, in the initial planning of MI-E device use, to start with low pressures and make adjustments to the pressure settings according to clinical parameters (29, 30).

Just as there is no consensus in the literature related to the settings of MI-E device use, there are also insufficient data related to the ideal durations of daily and weekly use. Meric et al. demonstrated in their study that MI-E device improved vital capacity immediately (31). This improvement was still present one hour after treatment and eventually reduced chest-wall motion asymmetry. However, since its benefits were lost rapidly, the machine should be used several times daily. As in the current study, previous reports have been related to use in clinical practice. In a small questionnaire study of 37 patients by Mahede et al., fewer than 50% of patients used the MI-E device daily (32). In the current study, the majority of the subjects used the device daily. The device was reported to be used a mean of 2.26 ± 1.46 times a day when the subject was not ill, and a mean of 3.67 ± 1.54 times a day when ill. It was considered that the young age of the subjects, that they were receiving non-invasive/invasive respiratory support, and that they had tracheostomies could have increased compliance with the device.

There were some limitations to this study, primarily that because of the cross-sectional design it was not possible to compare QoL scores before and after the device use in the NMD subjects. However, the control group was matched with respect to age, gender, and respiratory parameters. This was thought to be an important reason why no change could be detected in the QoL scores of all the groups and therefore, prospective studies are needed on this subject. Secondly, MI-E is an expensive device and there would be a bias that the study group has an economic advantage over the control group but almost all of the study patients obtained the device through a charity campaign and also we did not find any difference in the QoL-FRTP between the study and control groups. Finally, the data related to the daily and weekly use of the MI-E device was not recorded from the device but was taken as stated by the parents of the subjects.

CONCLUSION

The results of this study demonstrate that the use of an MI-E device made a positive contribution to ease of expectoration, number of hospital admissions, and length of stay in patients with NMD, and it also contributed positively to QoL in patients with SMA. Nevertheless, as there are insufficient data in the literature related to the ideal settings for the MI-E device, there is a need for further studies.

Ethics Committee Approval:
This research complies with all the relevant national regulations, institutional policies and is in accordance with the tenets of the Helsinki Declaration, and has been approved by the Akdeniz UniversityEthical Committee, Akdeniz University (Approval no KAEK-29; date 13/1/2021).

Informed Consent:
All the participants' rights were protected and written informed consents were obtained before the procedures according to the Helsinki Declaration.

Author Contributions:
Concept-A.E.B., A.B.; Data Collection and/or Processing- T. K, S. D, Ö. D., Ş. H.; Analysis and/ or Interpretation- A.E.B., A.B., A.B.; Writing Manuscript- A.E.B., A.B.; Critical Review- Ö.D., Ş.H., A.B.

Conflict of interest:
The authors have no conflict of interest to declare.

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