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Original Article

Rate of oral intake and effects of mechanical insufflation-exsufflation on pulmonary complications in patients with duchenne muscular dystrophy

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Abstract. [Purpose] In Duchenne muscular dystrophy, it increases risks of difficulties of expectoration of secretion, asphyxia, aspiration pneumonia because of decreased cough function. The aim of this study is to prove that manually assisted coughing or mechanical insufflation-exsufflation prevents pulmonary complication and contribute to continue oral intake safely and continue rate of oral intake in Duchenne muscular dystrophy. [Subjects and Methods] We investigated the status of using ventilator, manually assisted coughing or mechanical insufflation-exsufflation, and oral intake or not. In addition, we inspected the frequency of fever (over 37 °C) needed antibiotics from medical records for index of respiratory tract infection, and compared with every period of using mechanical insufflation-exsufflation from respiratory evaluation on cough peak flow. [Results] Fifty-eight patients participated in this study. There were 45 Full-time noninvasive positive pressure ventilation patients. Forty-three in 45 Full-time noninvasive positive pressure ventilation patients (95.6%) avoided tracheostomy and continued noninvasive positive pressure ventilation because they continued oral intake without tracheal intubation due to the respiratory acute exacerbation by asphyxia or aspiration pneumonia. [Conclusion] Duchenne muscular dystrophy patients can continue oral intake safely while preventing pulmonary complication by using manually assisted coughing or mechanical insufflation-exsufflation.

Key words: Duchenne muscular dystrophy (DMD), Mechanical insufflation-exsufflation (MI-E), Noninvasive positive pressure ventilation (NPPV)

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INTRODUCTION

Neuromuscular diseases (NMD), including Duchenne muscular dystrophy (DMD), cause ventilatory insufficiency due to weakness of inspiratory muscles, such as the diaphragm[1, 2)]. Loss of lung and chest wall compliance aggravates respiratory insufficiency. Moreover, loss of ventilatory muscle strength and laryngopharyngeal function may cause difficulty in expectoration of airway secretions and foreign substances, and acute respiratory failure due to asphyxia or aspiration pneumonia[3)].

Chewing and swallowing problems are common in NMD and reflect the weakness of muscles in the cheeks, lips, tongue and pharynx; these frequently contribute to malnutrition. Ingestion and swallowing difficulties may occur at a certain age in NMD patients. In spinal muscular atrophy, these can occur from infancy to childhood[4)], whereas in DMD, swallowing dif-

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ficulties usually happen after 15 years of age, following the appearance of respiratory failure. As a consequence of impaired oropharyngeal function, patients suffer from decreased quality of life (QOL)[5] and increased risk for aspiration pneumonia, which might contribute to or exacerbate respiratory failure[4].

In recent years, noninvasive positive pressure ventilation (NPPV) has been the first choice for respiratory management of NMD[6]. Airway clearance with manually assisted coughing or mechanical insufflation-exsufflation (MI-E) had been recommended by guidelines across Japan, Europe, and United States for patients with impaired cough function to prevent pulmonary complications of asphyxia or aspiration and to avoid tracheostomy or tracheal intubation[7–13]. In fact, these respiratory management strategies have been shown to significantly improve life expectancy and QOL of DMD patients[14, 15].

The aim of this study is to prove that manually assisted coughing or mechanical insufflation-exsufflation prevents pulmonary complication and contribute to continue oral intake safely and continue rate of oral intake in Duchenne muscular dystrophy.

SUBJECTS AND METHODS

The subject was a patient who was able to evaluate respiratory function in DMD patients hospitalized in our hospital at December 2014. DMD patients who could not undergo respiratory evaluation due to impaired comprehension, autism, and presence of tracheostomy were excluded.

We reviewed the medical records for variables, such as use of mechanical ventilator, use of MIE, and ability for oral intake. We investigated the duration of four periods of varying cough peak flow (CPF) value on unassisted coughing and manually assisted coughing and MI-E use (Fig. 1). CPF was measured using a low range peak flow meter (ASSESS; HealthScan, Inc., Cedar Grove, NJ), whereas MI-E was administered by CoughAssist and CoughAssist E70 (Philips Respironics, Inc., Murrysville, PA). This investigation was conducted with the approval of the ethics committee of our hospital (Registration number 28-1).

Period A: unassisted CPF ≥270 l/min before MI-E use.

Period B: unassisted CPF<270 l/min and manually assisted CPF ≥270 l/min before MI-E use.

Period C: unassisted CPF<270 l/min and manually assisted CPF<270 l/min with MI-E used for emergency only.

Period D: unassisted CPF<270 l/min and manually assisted CPF<270 l/min with constant use of MI-E.

We reviewed the medical records of each period for signs of respiratory tract infection, which was determined based on frequency of fever (temperature >37 °C) that required antibiotics. The variables were compared among the four periods using one-way analysis of variance with Bonferroni correction. The level of significance was set at a p value of <0.05. Statistics analysis was performed using SPSS Ver 22.0 J for Windows (IBM SPSS Japan Inc., Tokyo, Japan).

RESULTS

A total of 58 DMD patients participated in this study; their average age was 28.7 ± 8.0 (range, 14–45) years. Ventilatory support was not needed in 6 patients [age, 18.4 ± 2.7 (range, 15–23) years]; whereas nocturnal NPPV was needed in 7 patients [age, 21.4 ± 5.1 (range, 14–28) years] and full-time NPPV in 45 patients [age, 31.2 ± 7.0 (range, 17–45) years]. All patients who were not on ventilatory support and those on nocturnal NPPV were able to tolerate oral intake. Among the 45 patients on full-time NPPV, 43 (95.6%) were able to continue oral intake for an average of 7.2 ± 4.7 years (maximum 16.4 years), without the need for tracheal intubation due to acute respiratory failure from asphyxia or aspiration pneumonia (Table 1). The two remaining full-time NPPV patients could not tolerate oral intake and required nasogastric tube feeding due to frequent choking at 33.2 and 36.1 years old, respectively. Full-time NPPV patients was using NPPV during their meals in all cases.

In all full-time NPPV patients, MI-E was introduced when CPF was <270 l/min. In 35 of 45 patients, MI-E was used constantly at 3–5 times/day (Period D). In the remaining 10 of 45 patients, MI-E was unnecessary or was difficult to use continuously every day; instead, MI-E was used only for emergency situations, such as choking and difficulty in sputum clearance, at a frequency of 0–1 time/month (Period C).

In regards to the progression of CPF and MI-E usage, as well as the frequency of fever requiring antibiotics in each period, For periods A, B, C, and D, the cumulative number of patients was 15, 33, 12, and 37, respectively; the frequency of fever requiring antibiotics was 39, 91, 173, and 258, respectively (Table 2). Period C tended to have the highest rate of respiratory infection, but there was no statistically significant difference among the different periods.

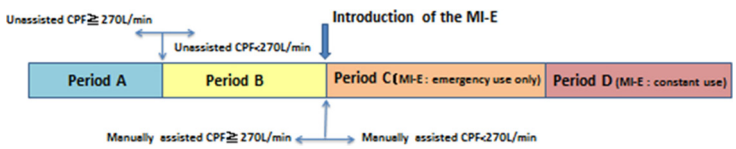

Fig. 1. Different study periods based in CPF value on unassisted coughing and manually assisted coughing and use of MI-E

DISCUSSION

In our study, 95.6% of the DMD patients on full-time NPPV were able to continue oral nutrition. A large-scale, multicenter study by Saitou et al. showed that despite the use of mechanical ventilation in a majority of hospitalized DMD patients, the rate of oral intake markedly decreased from 95.1% in 1999 to 66.8% in 2012[16]. On the other hand, Wollinsky reported that only 3 (14.3%) of 21 DMD patients using full-time NPPV required gastrostomy tube, whereas the remaining 18 (85.7%) were able to continue oral nutrition while using NPPV[17]. The high rate of oral intake in our study population may be explained by the fact that our patients continued full-time NPPV during their meals.

Servera et al. reported that an available MI-E at home was more certain than manually assisted coughing techniques (i.e., use of resuscitation bag and chest thrust) in managing asphyxia due to severe food aspiration in a 29 year-old DMD patient[18]. In our hospital, constant use of MI-E or the presence of a staff member who was knowledgeable in MI-E preparation during an emergency may explain the prolonged and safe oral intake of the majority of our patients on full-time NPPV.

The frequency of fever requiring antibiotics was the same between periods D and A (0.79 vs. 0.76, respectively) and there was no clear episode of aspiration pneumonia. Bianch compared the data of CPF and other respiratory function tests with and without respiratory complications such as repeated bronchitis and pneumonia at the convalescent stage when restarting oral intake in 55 patients that had cerebrovascular disorder or neurological disorder, with swallowing difficulty confirmed by video fluoroscopic examination (VF). The results showed that the CPF value of patients with respiratory complications was significantly lower than those without complications, and complications emerged when the value was under 242 l/min[19].

Toussaint described that use of daytime NPPV may help DMD patients with dysphagia in recovering from dyspnea or respiratory muscle fatigue during a meal. They recommended respiratory management using NPPV and as needed manually 4assisted coughing and noninvasive airway clearance by MI-E to avoid tracheostomy in DMD patients with dysphagia[5].

At our hospital, the setting for MI-E was usually at +40 to +55 cmH_2O positive pressure and −40 to −55 cmH_2O negative pressure[20, 21], and was adjusted to gain enough CPF of ≥270 l/min). In this study, all patients on full-time NPPV were noted to have improved cough function from an unassisted CPF of 54.1 ± 62.6 l/min to CPF>270 l/min after use of MI-E. We considered that the use of MI-E and alteration of food consistency (pasty or minced) in these patients were able to prevent pulmonary complications, despite choking or wheezing during a meal.

Table 1. Characteristic of all patients at the end of study

		No ventilatory support (n=6)	Nocturnal NPPV (n=7)	Full-time NPPV (n=45)
Age (range)		18.4 ± 27 (15–23)	21.4 ± 5.1 (14–28)	31.2 ± 7.0 (17–45)
VC (ml)		2,081.7 ± 602.1	1,118.6 ± 687.3	287.4 ± 211.9
MIC (ml)		2,537.5 ± 179.7	2,311.4 ± 510.1	1,703.0 ± 708.7
CPF (L/min)		294.2 ± 71.0	202.1 ± 96.7	54.1 ± 62.6
assisted CPF (L/min)		356.7 ± 51.3	324.2 ± 58.6	254.2 ± 70.3
MI-E	Constant use	0 (0%)	2 (28.6%)	35 (77.8%)
	Emergency use only	6 (100%)	5 (71.4%)	10 (22.2%)
Power-operated vehicle	enable	6 (100%)	7 (100%)	42 (93.3%)
	disable	0 (0%)	0 (0%)	3 (6.7%)
Nutrition method	Oral feeding	6 (100%)	7 (100%)	43 (95.6%)
	Tube feeding	0 (0%)	0 (0%)	2 (4.4%)

NPPV: noninvasive positive pressure ventilation; VC: vital capacity; MIC: maximum insufflation capacity; CPF: cough peak flow; assisted CPF: manually assisted cough peak flow: MI-E: mechanical insufflation-exsufflation

Table 2. Frequency of fever needed antibiotics treatment as respiratory tract infections

	Total patients	Times of RTIs	Patients years	RTIs/ patients-years	RTI/year/ patient	
Period A	15	39	59.8	0.65	0.76	N, S
Period B	33	91	148.9	0.61	0.67	
Period C	12	173	100.1	1.73	1.35	
Period D	37	258	308.3	0.84	0.79	

RTIs: respiratory tract infections

One limitation of this study was that we were unable to ascertain whether the febrile episodes that required antibiotics were from aspiration pneumonia or from other respiratory tract infections. The diagnosis of ingestion-swallowing difficulty in patients on full-time NPPV was not confirmed by video fluoroscopy (VF). However, VF is not always useful and an ingestion-swallowing difficulty may be easily observed during a meal and based on changes in ventilation[5, 22].

Use of manually assisted coughing and noninvasive airway clearance by MI-E enabled continuous and safe oral intake in DMD patients on full-time NPPV. Both NPPV and MI-E are important in avoiding tracheal intubation or tracheostomy and might improve the life expectancy and QOL of DMD patients.

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