Effects of Positioning on Cough Peak Flow and Muscular Electromyographic Activation in Duchenne Muscular Dystrophy

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BACKGROUND: Advanced stages of Duchenne muscular dystrophy (DMD) result in muscle weakness and the inability to generate an effective cough. Several factors influence the effectiveness of cough in patients with DMD. The aim of this study was to assess whether differences in positioning affect cough peak flow (CPF) and muscular electromyographic activation in subjects with DMD compared with paired healthy subjects. **METHODS:** Optoelectronic plethysmography and surface electromyography were used to assess chest wall volumes, chest wall inspiratory capacity, CPF, breathing pattern, and electromyographic activity of sternocleidomastoid, scalene, rectus abdominis, and external oblique muscles during inspiratory and expiratory cough phases in the supine position, supine position with headrest raised at 45°, and sitting with back support at 80° in 12 subjects with DMD and 12 healthy subjects. **RESULTS:** Subjects with DMD had lower CPF ($P < .01$) in comparison to control subjects in all positions; the DMD group also exhibited lower CPF ($P = .045$) in the supine position versus 80°. Moreover, the relative volume contributions of the rib cage and abdominal compartments to tidal volume modified significantly with posture. The electromyographic activity during inspiratory and expiratory cough phases was lower in subjects with DMD compared to healthy subjects for all evaluated muscles ($P < .05$), but no significant differences were observed with posture change. **CONCLUSIONS:** In subjects with DMD, posture influenced CPF and the relative contribution of the rib cage and abdominal compartments to tidal volume. However, muscular electromyographic activation was not influenced by posture in subjects with DMD and healthy subjects. *Key words:* Duchenne muscular dystrophy; cough; plethysmography; respiratory muscles; electromyography. [Respir Care 2020;65(11):1668–1677. © 2020 Daedalus Enterprises]

Introduction

Duchenne muscular dystrophy (DMD) is an X-linked neuromuscular disease resulting in progressive weakness

and wasting of all the striated muscles, including the respiratory muscles, with an incidence of one in 3,500 live male births.^{1,2} Up to now, respiratory problems in DMD have been described mainly in terms of reduction of vital

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capacity and inspiratory muscle weakness. In DMD, the linear decline of FVC expressed as a percentage of the predicted values through the years indicates progressive respiratory muscle weakness.^{3,4}

With the progression of the disease and worsening of respiratory muscles function, cough, whose function is to maintain airway clearance and pulmonary hygiene, becomes critical.⁵⁻⁷ In DMD, ineffective cough leads to severe respiratory complications, such as ineffective airway clearance, pneumonia, atelectasis, and intubations.⁵ Several factors may lead to ineffective coughing, such as structural changes of the chest wall, reduced precough inspiratory volume, and respiratory muscle weakness.⁸⁻¹⁰ This weakness prevents the patient from taking a deep breath, decreasing the volume during the expiratory coughing phase, with aggravation in the presence of expiratory musculature impairment.^{9,11,12}

Positioning leads to changes in thoracoabdominal kinematics during quiet spontaneous breathing in individuals with DMD.^{4,13} With the advancement of the disease, there is a significant and progressive reduction of the abdominal contribution to quiet spontaneous breathing while supine. According to LoMauro et al,⁴ the chest wall motion during spontaneous breathing is an important indicator of the degree of respiratory muscle impairment. However, the behavior of respiratory muscle activation during coughing in different positions is still unknown in patients with DMD as well as its influence on cough peak flow (CPF). The objective of this study was to assess whether differences in positioning affect CPF and muscular electromyographic activation in subjects with DMD compared with age-matched healthy subjects. We hypothesized that the positioning would influence muscular electromyographic activation and CPF generation, which would be stronger when the torso was more erect due to changes in the thoracoabdominal kinematics and muscular activation.

Methods

Subjects and Inclusion Criteria

This cross-sectional study was carried out according to the guidelines of the Helsinki Declaration and was

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QUICK LOOK

Current knowledge

In patients with Duchenne muscular dystrophy, breathing patterns and kinematics of the chest wall are influenced by age and disease during spontaneous breathing with a predominance of the thoracic compartment over the abdominal in the supine position. Optoelectronic plethysmography allows a real-time assessment of a natural cough and chest wall volumes and avoids interference in the regular breathing pattern.

What this paper contributes to our knowledge

The cough peak flow is influenced by positioning in individuals with Duchenne muscular dystrophy. Respiratory muscle activation is not affected by different positions. The study also reveals that this population presented lower values of cough peak flow and muscle activation compared to control subjects. In addition, muscular activation must be taken into account when monitoring disease progression.

approved by the Research Ethics Committee of the University Hospital Onofre Lopes. All children in the study (when possible) and the legal guardian of each child signed the free, informed consent after explanation of all procedures, as proposed and approved by the institution's Commission of Ethics and Research.

The DMD group consisted of 12 subjects > 7 y old with a confirmed clinical diagnosis of DMD evaluated by a neurologist based on traditional diagnostic criteria: progressive muscular deficit resulting in severe motor disability; increased muscle plasma enzymes; muscle biopsy identifying muscular degeneration and absence of dystrophin; alterations in the DMD gene (eg, deletions, duplications, or point mutations),¹⁴ with no associated infectious pulmonary diseases, no tracheostomy, and no use of sedatives or non-invasive ventilation for > 16 h/d. Subjects with DMD who did not complete the evaluation or were unable to adhere to the study protocol were excluded. The control group consisted of 12 male subjects paired according to age who reported themselves to be healthy without any history of cardiovascular, restrictive, or obstructive pulmonary disease. Subjects who presented values < 80% of predicted FVC or FEV₁, as well as those who failed to perform the study protocol, were excluded.

Pulmonary Function

Pulmonary function was assessed using a KoKo Digi-Doser spirometer (nSpire Health, Longmont, Colorado) according to the protocol recommended by the American

Thoracic Society/European Respiratory Society.¹⁵ The predicted values were calculated as described by Quanjer et al.¹⁶ The highest values of each test were included in the statistical analysis.

Respiratory Muscle Strength

Respiratory muscle strength was evaluated using a digital manometer (NEPEB-LabCare, Belo Horizonte, Brazil) to measure maximum inspiratory and expiratory pressures from residual volume and total lung capacity, respectively (see supplementary materials at <http://www.rcjournal.com>).^{15,17} The sniff nasal inspiratory pressure test also evaluated inspiratory muscle strength. The predicted values for all respiratory muscle strength assessments were calculated using the reference values proposed for individuals 7–18 y old and for individuals > 18 y old.^{18–21} The highest values of each test were included in the statistical analysis.

Chest Wall and Compartmental Volumes and Cough Peak Flow

An optoelectronic plethysmography (OEP) system (BTS Bioengineering, Milan, Italy) was used to assess total chest wall and compartmental (ie, pulmonary rib cage, abdominal rib cage, and abdomen) volumes, breathing pattern, and CPF.^{22,23} The measurement of CPF with OEP is a validated method.^{10,24–26} The analyses were carried out with each subject positioned in a hard bed in the supine position (180°), the supine position with headrest raised at 45°, sitting with back support at 80° (80°), and seated without back support due to muscle weakness. To calculate the volume changes of the posterior part of the trunk (hidden by the bed support), a virtual plane was defined and obtained by calculating a reference plane with the coordinates of the markers positioned laterally on the trunk of the individuals.²⁷

For this study, the pulmonary rib cage and the abdominal rib cage compartments were considered as a single compartment (ie, rib cage), given by their sum.^{28–31} The analyses of the following variables were performed based on data obtained from the OEP: CPF derived from the displacement of the chest wall volume over time during coughing^{28–31}; inspiratory capacity of the chest wall, the rib cage, and the abdomen during cough; the percentage of contribution of the rib cage and abdomen to the inspiratory capacity; volume of expired air from the chest wall during coughing; change in chest wall tidal volume and its compartments (ie, rib cage and abdomen); the percentage of contribution of the compartments to the change in tidal volume (ie, rib cage and abdomen); inspiratory time (T_I), expiratory time (T_E), and total respiratory cycle time (T_{tot}); the percentage of inspiratory time in relation to total time (T_I/T_{tot}); breathing frequency (f); expiratory minute volume ($\dot{V}_E$); and the rapid shallow breathing index (RSBI), calculated as $f/\dot{V}_T$.³²

Fig. 1. Representative tracings of chest wall volume (V_{CW}), flow, and raw SCM, scalene, rectus abdominis and external oblique muscles in one healthy subject and one Duchenne muscular dystrophy subject during a cough maneuver. CPF = cough peak flow; V_{CW} = chest wall volume; SCM = signals of the sternocleidomastoid.

Electromyographic Recordings

A TeleMyo DTS Desk Receiver electromyograph was used with 4 wireless Clinical DTS sensors (Noraxon USA, Scottsdale, Arizona) for surface electromyographic signal acquisition and processing. Signal acquisition was performed at a sampling frequency of 1,500 Hz with a 500-Hz low-pass filter, a gain of 1,000 times, and a common mode rejection index > 120 dB. All data were analyzed using MR 3.8 software (Noraxon USA, Scottsdale, Arizona). Electromyographic data acquisition was performed synchronously with OEP (Fig. 1).

The passive bipolar surface electrodes were set according to SENIAM³³ recommendations on the following muscles: sternocleidomastoid, in the lower third of the

distance between the mastoid process and the sternoclavicular joint³⁴; scalene, at 5 cm from the sternoclavicular joint and 2 cm above this point³⁵; rectus abdominis at 4 cm from the umbilical scar³⁴; and external oblique, at 50% of the upper anterior iliac spine line and the tip of the eleventh rib.³⁵ All electrodes were placed on the right side of the body to reduce cardiac signal noises. The electromyographic signals were standardized as a percentage of root mean square. The signal was processed by applying the 20-Hz high-pass filter, full wave rectification, smoothing with the root mean square algorithm and a 50-ms window, and a 30-Hz Butterworth high-pass filter. All signals were normalized from the average of 3 maximum voluntary isometric contractions for the sternocleidomastoid, scalene, rectus abdominis, and external oblique; values were standardized as being equal to 100%.³⁴⁻³⁶

The fold change of the muscles during the inspiratory and expiratory phases was calculated by subtracting the normalized activation value of the evaluated muscle in spontaneous breathing (B) from the normalized muscle activation value in the evaluated cough stage (A) (ie, inspiratory or expiratory) in the same position divided by (B): fold change = (A – B)/B. This calculation indicates the number of times the electromyographic signal increased or decreased from baseline.

Functional Scale

The Motor Function Measurement scale³⁷ was used to assess functionality in subjects with DMD. This scale consisted of 32 items divided into 3 dimensions: standing position and transfers (13 items), axial and proximal motor function (12 items), and distal motor function (7 items). Each item is graded from 0 to 3 points. The total score and scores in each domain are given as percentages; the higher the score, the better the motor function.

Study Design

After acquiring anthropometric, pulmonary function, and respiratory muscle strength data, subjects were assessed on a standard bed at 3 different positions randomized by simple draw to perform the tests at 180°, 45°, and 80° with 10 min rest time between positions. In all positions, OEP data were recorded in 3 phases: phase 1 involved 60 s of quiet spontaneous breathing; phase 2 involved 2 slow vital capacity maneuvers with 30 s of quiet spontaneous breathing in between; and phase 3 involved 3 spontaneous cough maneuvers (with the expiratory phase initiated from the highest inspiratory capacity reached) with 30 s of quiet spontaneous breathing in between (see the figure in the supplementary materials at <http://www.rcjournal.com>). For each subject, the highest CPF was considered for data analysis.

Statistical Analysis

For sample size calculation, CPF was assessed in a pilot study conducted with 4 subjects with DMD (2 subjects were 13 y old, 1 subject was 17 y old, and 1 subject was 20 y old) and 4 age-matched control subjects in all positions. With a η_p^2 of 0.687, an effect-size f of 1.48, an alpha error of 0.05, and a statistical power of 95%, a total sample of 20 individuals (ie, 10 subjects in each group) was estimated.

Data are expressed as mean $\pm$ SD unless otherwise stated. Data normality and distribution were performed using the Shapiro-Wilk test. Intergroup comparisons regarding anthropometric data, lung function, and respiratory muscle strength were performed using the unpaired t test. Intergroup analyses regarding OEP and surface electromyographic data were compared using the Mann-Whitney test, whereas intragroup analyses were performed using the Kruskal-Wallis test with the Dunn post hoc test. Intragroup (r) and intergroup (ϵ^2) nonparametric effect sizes were calculated according to Fritz et al³⁸ and Tomczak and Tomczak,³⁹ respectively (see supplementary tables at <http://www.rcjournal.com>). For the former, values were interpreted as small (< 0.10), moderate (between 0.10 and 0.30), and large (> 0.50)³⁸; for the latter, values were interpreted as small (< 0.06), moderate (between 0.06 and 0.14), and large (> 0.14).³⁹

A significance level of $P < .05$ was adopted. All statistical procedures were performed using Prism 6.0 for Windows (GraphPad, San Diego, California), and the sample calculation and effect sizes were performed using Gpower 3.1.9.2 (Kiel, Germany).

Results

Sample Characteristics

Anthropometric data were only statistically different for height ($P = .01$), which was lower in subjects with DMD when compared to the control group. Predictive and absolute values of lung function and respiratory muscle strength were significantly lower (all $P < .001$) in subjects with DMD when compared to controls, except for FEV₁/FVC ratio. Subjects with DMD had more preserved distal motor function than foot and transfer function, and axial and proximal motor function (Table 1).

Chest Wall Compartmental Volumes and Breathing Pattern

During quiet spontaneous breathing, subjects with DMD presented lower changes in chest wall tidal volume ($P = .044$) and lower changes in tidal volume in the rib cage compartment ($P = .025$) in the supine position and the 45° position, respectively, compared to controls (Table 2). A

Table 1. Subject Characteristics

	Control Group	Duchenne Group	<i>P</i>
Age, y	15.17 ± 5.35	15.33 ± 5.49	.99
Height, m	1.57 ± 0.16	1.46 ± 0.14	.01
Weight, kg	52.44 ± 2.58	48.58 ± 16.29	.50
Body mass index, kg/m ²	20.44 ± 5.03	22.10 ± 5.11	.61
FVC, L	3.36 ± 1.32	1.76 ± 0.66	.001
FVC, % of predicted	99 ± 15.38	60.3 ± 16.8	< .001
FVC, <i>z</i> score	-0.07 ± 1.20	1.19 ± 2.42	< .001
FEV ₁ , L	2.84 ± 1.06	1.44 ± 0.74	.001
FEV ₁ , % of predicted	97.8 ± 14.1	53.4 ± 17.8	< .001
FEV ₁ , <i>z</i> score	-0.16 ± 1.20	-3.88 ± 1.47	< .001
FEV ₁ /FVC	0.85 ± 0.07	0.78 ± 0.16	.15
FEV ₁ /FVC, <i>z</i> score	-0.10 ± 1.40	-3.65 ± 1.58	.23
FEF _{25-75%} , L/s	3.27 ± 1.02	1.86 ± 1.22	.006
FEF _{25-75%} , %	99.8 ± 16.7	55.7 ± 28.7	< .001
FEF _{25-75%} , <i>z</i> score	-0.03 ± 0.74	1.71 ± .49	.002
P _{imax} , cm H ₂ O	111.1 ± 25.8	59.25 ± 18.66	< .001
P _{imax} , % of predicted	110.4 ± 26.3	55.5 ± 17.9	< .001
P _{emax} , cm H ₂ O	122.4 ± 31	51.83 ± 14.70	< .001
P _{emax} , % of predicted	114.4 ± 25.6	44 ± 12.1	< .001
SNIP, cm H ₂ O	91.83 ± 13.47	54.3 ± 15.57	< .001
SNIP, % of predicted	79.2 ± 13.9	47.7 ± 15.7	< .001
Motor function measurement, % total	NA	40.25 (33.5–82.7)	NA
Motor function measurement, % of D1	NA	0 (0–59.7)	NA
Motor function measurement, % of D2	NA	55 (42.5–98.5)	NA
Motor function measurement, % of D3	NA	9.3 (55.7–100)	NA
Wheelchair bound	NA	8 (66)	NA
Noninvasive mechanical ventilation	NA	3 (25)	NA
Use of cough-assist devices	NA	0	NA
Steroid treatment	NA	12 (100)	NA

Data are presented as *n* (%), mean ± SD, or median (interquartile range). Both groups had 12 subjects.

NA = not applicable; P_{imax} = maximum inspiratory pressure; P_{emax} = maximum expiratory pressure; SNIP = sniff nasal inspiratory pressure; D1 = Domain 1, standing and transfers; D2 = Domain 2, axial and proximal motor function; D3 = Domain 3, distal motor function

higher *f* ($P < .001$), lower T_I ($P = .03$), and lower T_E ($P < .001$) were observed in all positions in the DMD group compared to the control group. Subjects with DMD also exhibited lower T_{tot} ($P = .007$) and a higher T_I/T_{tot} ($P = .001$) in the supine position compared to the respective position of the control group (Table 3). The RSBI was higher in subjects with DMD at all positions when compared to controls (Table 3).

Regarding the intragroup analysis, subjects with DMD presented lower changes in tidal volume contribution by the rib cage in supine position ($P < .05$) and higher changes in tidal volume contribution by the abdomen in the supine and 45° positions ($P < .05$) compared to the 80° position (Table 3). A higher T_I/T_{tot} ($P = .01$) was also observed in the supine position compared to the 80° position (Table 3).

Cough Peak Flow

CPF was lower in subjects with DMD when compared to controls in all positions: supine ($P < .001$), 45° ($P =$

.001), and 80° ($P < .001$). Regarding intragroup analysis, a lower CPF ($P = .045$) was observed in subjects with DMD in the supine position when compared to the 80° position (Fig. 2).

Inspiratory Capacity and Variation in Expiratory Air Volume During Coughing

The DMD group exhibited lower chest wall inspiratory capacity ($P = .02$) and rib cage inspiratory capacity ($P = .02$) in comparison to the control group in all positions. The abdominal inspiratory capacity was also lower in the DMD group at 80° ($P = .01$) when compared to controls. Subjects with DMD also exhibited a lower variation in the chest wall expiratory volume during coughing in all positions ($P < .05$) in the DMD group compared to the respective positions of the control group (Table 2). Regarding intragroup analysis, DMD ($P = .01$) and controls ($P = .02$) presented lower rib cage inspiratory capacity in the supine position compared to the 80° position (Table 2).

Table 2. Effects of Position on Chest Wall Compartmental Tidal Volume and Inspiratory Capacity During Cough

	Control Group				Duchenne Group			
	Supine		45°		Supine		45°	
	Intragroup	P	Intragroup	P	Intragroup	P	Intragroup	P
ΔTidal volume, L								
Chest wall	0.33 (0.24–0.52)	0.39 (0.23–0.56)	0.35 (0.19–0.60)	.95	0.22 (0.29–0.18)*	.02	0.21 (0.18–0.28)*	.044
Rib cage	0.13 (0.10–0.19)	0.15 (0.10–0.27)	0.19 (0.11–0.33)	.55	0.08 (0.06–0.13)*	.02	0.09 (0.60–0.14)*	.02
Abdomen	0.17 (0.09–0.31)	0.19 (0.13–0.29)	0.13 (0.07–0.22)	.30	0.13 (0.09–0.16)	.17	0.13 (0.09–0.15)	.13
ΔTidal volume, %								
Rib cage	47.0 (37–58)	46.5 (4.6–5)	59.0 (52.5–64.7)	.061	42.0 (22.2–57.5)†	.43	41.5 (24.0–54.5)	.50
Abdomen	53.5 (42.2–63.7)	53.8 (44.7–59.5)	41 (35.9–5.7)	.063	58.5 (42.5–77.7)†	.48	59.0 (46.2–76.2)†	.54
Inspiratory capacity, L								
Chest wall	2.03 (1.19–3.20)	2.09 (1.31–2.97)	2.99 (1.93–3.12)	.31	1.27 (0.85–1.44)*	.02	1.21 (0.84–1.53)*	.008
Rib cage	1.49 (0.09–2.10)†	1.70 (1.11–1.98)	2.23 (1.47–2.48)	.02	0.90 (0.66–0.96)*†	.006	1.02 (0.76–1.10)*	.008
Abdomen	0.43 (0.33–1.09)	0.46 (0.23–1.02)	0.63 (0.34–0.78)	.83	0.43 (1.23–0.54)	.28	0.21 (0.05–0.50)*	.07
Inspiratory capacity, %								
Rib cage	73.0 (64.7–79.7)	73.0 (65.5–85.2)	77.5 (73.1–86.5)	.30	73.6 (61.0–84.1)	.80	84.7 (66.6–97.0)	.33
Abdomen	28.0 (21–35.5)	27.0 (15.5–34.9)	22.6 (13.7–27.2)	.26	27.1 (16.3–9.7)	.83	16.2 (5.2–34.5)	.38
ΔVolume of expired air	1.70 (1.16–2.84)	2.10 (1.16–3.23)	2.7 (1.8–3.25)	.005	0.90 (0.65–1.34)	.21	1.03 (0.73–1.66)	.01

Data are presented as median (interquartile range).

* Intergroup difference with the respective position of the control group. Negative values during the inspiratory capacity maneuver are due to paradoxical movement of the abdominal compartment. Neither the degree of dyssynchrony between compartments nor the paradoxical movement was assessed in this study.

† Intragroup difference with position 80°.

Electromyographic Recordings

Subjects with DMD exhibited significantly lower inspiratory muscle activation (sternocleidomastoid and scalene) during the inspiratory coughing phase ($P < .05$) compared to the control group in all positions; the DMD group was more expressive at the 45° position (Fig. 3). The expiratory muscles were only slightly activated in the subjects with DMD compared to control subjects at 45° and 80° ($P < .05$) for rectus abdominis, and in the supine ($P = .050$) and 80° ($P = .001$) positions for external oblique (Fig. 2). Subjects with DMD presented less muscle activation ($P < .05$) for all evaluated muscles compared to control subjects during the expiratory coughing phase (Fig. 3). No intragroup differences were observed in both groups during the coughing phases.

Discussion

To our knowledge, this is the first study to combine the use of OEP to assess voluntary CPF and surface electromyographic activation of the respiratory muscles in subjects with DMD compared to healthy subjects in different positions. The main results of this study were that, when compared to paired healthy subjects, subjects with DMD exhibited higher CPF at 80° compared to the supine position and lower CPF and respiratory muscle activation values in all positions.

Previous studies reported that, in subjects with DMD, breathing pattern and kinematics of the chest wall are influenced by age and disease during spontaneous breathing with a predominance of the thoracic compartment over the abdominal compartment in the supine position.^{4,13} However, this pattern is expected only after 14 y of age.⁴ In healthy subjects, conversely, the contribution of the thoracic compartment is mainly observed in the seated position, whereas the abdominal compartment is observed in the supine position.¹³ In our study, subjects with DMD presented a higher contribution of the abdominal compartment in both supine and 45° positions, similar to healthy subjects, because only 5 subjects in the DMD group were > 14 y old. However, the absolute abdominal volume in the DMD group was lower compared to the control group.

The supine position imposes mechanical changes on the chest wall, such as cranial diaphragm displacement, expiratory muscle stretching, and reduced lung volume and abdominal

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Fig. 3. Muscle activation during the inspiratory and expiratory phases of cough. Data are presented as average and standard error. Fold change (ie, A-B/B) of the electromyographic activation of the sternocleidomastoid muscle. SCM = sternocleidomastoid, DMD = Duchenne muscular dystrophy.

contribution to generate more volume to counterbalance the disadvantages of positioning and diaphragmatic weakness in subjects with DMD.^{4,10,49}

Our results during quiet spontaneous breathing indicate that subjects with DMD presented lower chest wall tidal volume at the expense of a higher f , resulting in a higher RSBI compared with controls, as a strategy to reduce the

elastic work within the breath and the perception of dyspnea.^{10,50} Our results agree with those reported by Misuri et al,⁵⁰ who evaluated subjects with neuromuscular disease using a pneumotachograph, including subjects with DMD, and observed that the subjects' ventilatory pattern showed a normal $\dot{V}_E$, a consistent decrease in T_I and T_E , and an increase in f and RSBI.

The DMD group exhibited lower muscle activation in all positions compared with healthy subjects. We also observed that the sternocleidomastoid and scalene inspiratory muscles in the control group extended their activation to the expiratory coughing phase. According to previous observations,⁵¹⁻⁵³ there is co-activation of the respiratory muscles to control precough inspiratory volume, flow, and pressure and to avoid distortion of the chest wall compartments due to the increased muscular activation during a normal cough maneuver.⁵⁴ In contrast, no co-activation of the inspiratory and expiratory muscles during the 2 phases of coughing was observed in subjects with DMD, probably due to a weakness of the respiratory muscles and consequently low CPF generation. Future studies could confirm the changes in co-activation of respiratory muscles as a new marker of disease progression.

Study Strengths and Limitations

To ensure the naturalness of a cough maneuver and to avoid interference (ie, mouthpieces, nose clips, or pneumotachograph) in the regular breathing pattern, we used the OEP system.⁵⁵ We believe that the OEP, which we consider to be a strength of our study, is particularly important to assess CPF in neuromuscular subjects because they frequently present problems in performing standard spirometric and $P_{\text{imax}}/P_{\text{emax}}$ tests due to facial weakness and consequent difficulty in keeping the mouthpiece in place.⁵⁶

Our study is limited by the sample size of subjects, which is related to a limited budget and time constraints. For this reason, subgroup analyses between noninvasive ventilation users and nonusers and between cough-assist device users and nonusers were not available. In addition, we did not evaluate the electromyographic signal of the diaphragm, which is the most responsible for the inspiratory phase of coughing and is likely to be among the most affected by position changes. Moreover, the expiratory activity of the sternocleidomastoid and scalene observed in the control group could represent a tendency to lean the neck forward during cough, rather respiratory compensation. For this reason, we tried to minimize this effect by stabilizing each subject's head and by instructing each subject to avoid flexing the neck during the cough. In addition, we did not evaluate body weight distribution, so we did not quantify the influence of central adiposity on respiratory mechanics.

Conclusions

In subjects with DMD, positioning influences the generation of CPF with no differences in respiratory muscle activation. When compared to age-matched healthy subjects,

subjects with DMD presented lower CPF values and less electromyographic activation during different coughing stages.

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