Respiratory abdominal muscle recruitment and chest wall motion in myotonic muscular dystrophy

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Ugalde, Viviane, Sandra Walsh, R. Ted Abresch, H. William Bonekat, and Eileen Breslin. Respiratory abdominal muscle recruitment and chest wall motion in myotonic muscular dystrophy. J Appl Physiol 91: 395–407, 2001.—Abdominal muscles are selectively active in normal subjects during stress and may increase the potential energy for inspiration by reducing the end-expiratory lung volume (EELV). We hypothesized that a similar process would occur in subjects with myotonic muscular dystrophy (MMD), but would be less effective, because of to their weakness and altered chest wall mechanics. Fine-wire electromyography (EMG) of the transversus abdominis (TA), internal oblique (IO), external oblique, and rectus abdominis was recorded in 10 MMD and 10 control subjects. EMG activity, respiratory inductive plethysmography, and gastric pressure were recorded during static pressure measurement and at increasing levels of inspiratory resistance breathing. EELV was reduced and chest wall motion was synchronous only in controls. Although the TA and IO were selectively recruited in both groups, EMG activity of the MMD group was twice that of controls at the same inspiratory pressure. In MMD subjects with mildly reduced forced vital capacity, significant differences can be seen in abdominal muscle recruitment, wall motion, work of breathing, and ventilatory parameters.

PULMONARY COMPLICATIONS ARE a common source of morbidity and mortality in myotonic muscular dystrophy (MMD) (7, 32). Respiratory muscle weakness, altered central ventilatory control, and impaired ventilatory mechanics lead to global alveolar hypoventilation, microatelectasis, and reductions in compliance (5, 17, 53). The presence of inspiratory respiratory muscle weakness in MMD is well established, with reductions in transdiaphragmatic pressure (43), pleural pressure (15), compound muscle action potential amplitude of the diaphragm (53), and maximal inspiratory pressure (MIP) (7, 31, 32, 43, 53). However, the first indication of pulmonary dysfunction in the MMD population is the decline in maximal expiratory pressure (MEP) (7, 32, 43). Whereas traditionally inspiratory muscle weakness has been the primary cause of respiratory failure, expiratory muscle dysfunction has recently been implicated in respiratory failure (26). Despite early declines in expiratory muscle strength, there has been little study of expiratory muscle action and corresponding abdominal wall mechanics in MMD.

Until recently, knowledge of abdominal respiratory muscle activation during respiration and their function in normal subjects has been limited. Abdominal muscles were thought to contract as a unit for expiratory accessory muscle action (24). Recent studies have shown that transversus abdominis (TA) and the internal oblique (IO) muscles are selectively active in normal subjects during stimulated ventilation or exercise (1, 2, 16, 47, 48). Expiratory recruitment of abdominal muscles theoretically enhances inspiration by causing passive abdominal recoil during initial inspiration, which aids descent of the diaphragm (19, 36). An objective, indirect measure of stored potential energy for inspiration is the reduction in end-expiratory lung volume (EELV) as seen during exercise and with pursed-lip breathing (28, 44, 47).

In addition to expiratory activity, the TA and IO are also recruited during inspiration in normal subjects during stimulated ventilation trials (1, 2, 48). Inspiratory abdominal activity may act as a counterforce to diaphragm descent optimizing the length-tension relationship in the area of apposition (18). Others postulate that the inspiratory abdominal activity may act as fine tuning of abdominal pressure and compliance and diaphragmatic descent (2) or as a braking mechanism to prevent excessive diaphragmatic descent with hyperinflation (24, 48).

Abdominal muscles are also known postural stabilizers in the erect position and are key for trunk motion (13, 14, 21, 29, 45). The abdominal muscle activity during stabilization is affected by respiration (29), and during respiration in the supine position abdominal muscle activity is minimal (1, 2).

Abdominal muscle activation and the mechanical response of chest wall motion in MMD subjects are less

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well understood. Selective TA and IO activity was seen during pursed-lip breathing in MMD subjects in a supine position with corresponding mechanical responses of increased gastric pressure and decreased abdominal volume (47). In a group of MMD subjects, abdominal paradox and chaotic breathing patterns were associated with decreased tidal volume and increased respiratory rate. With progressive increases in carbon dioxide rebreathing, the same group of MMD subjects demonstrated lower minute ventilation and lower tidal volume. With the reduced ventilatory responses, transient occlusion mouth pressure measurements were normal. Serisier et al. (43) hypothesized that the reduction in respiratory muscle strength in MMD subjects leads to disordered pulmonary mechanics, which result in greater inspiratory pressure needed to produce the same ventilation.

To date there have been no studies performed in the MMD population describing abdominal muscle recruitment patterns and corresponding mechanical changes in chest wall motion during exercise. Therefore, we conducted a controlled, observational study to determine the abdominal muscle recruitment during static maximal pressure measurements and during the exercise produced by inspiratory resistance breathing (IRB) and the subsequent effect of this recruitment on ventilation and chest wall mechanics. We also wished to study whether these changes resulted in an inspiratory biomechanical advantage, e.g., a reduction in EELV. We hypothesized the following. 1) In MMD subjects, the TA and IO would demonstrate selective activation of the abdominal muscles during static pressure measurements and during IRB. 2) MMD subjects would recruit the abdominal muscles at lower levels of inspiratory resistance than controls. 3) Mechanical responses of increased gastric pressure and the relative reduction in abdominal volume would correspond with phasic expiratory EMG activity of the abdominal muscles. 4) Corresponding to the expiratory activation of the abdominal muscles, a reduction in EELV would be seen. 5) MMD subjects would exhibit asynchronous breathing patterns when challenged with IRB in the supine position.

METHODS

Subjects

Ten ambulatory MMD subjects from a university-based neuromuscular disease clinic and ten normal controls voluntarily agreed to participate in this clinical investigation. The diagnosis of MMD was based on criteria described by Harper (27). The Human Subjects Review Board at our institution approved the study, and all participants signed a written consent form. All subjects were living at home, either independently or with assistive devices, were ambulatory at a limited household level, had no clinically significant comorbid medical or pulmonary problems, and were not taking any medications.

Subject Preparation

Measurements of pulmonary function (3), MIP, and MEP (8) were obtained and experimental trials of breathing against increasing inspiratory resistance were carried out by each subject. Parameters measured during these trials were fine-wire electromyography (EMG) of four abdominal muscles [TA, IO, external oblique (EO), and rectus abdominis (RA) muscles], gastric pressure, respiratory inductive plethysmography, mouth pressure, oxygen saturation, and Borg scale measurements of dyspnea, work of breathing, and fatigue.

Gastric Pressure

A Jaeger latex balloon catheter (Med Point Technologies, Stonyridge, OH) was inserted transnasally into the stomach, attached to a TSD104 pressure transducer (BioPac Systems, Goleta, CA) that was connected to a DA100 amplifier (BioPac Systems). Subjects were placed in the supine position with approximately a 15° angle at the waist for the remainder of the study. This position was chosen for two reasons. First, this position eliminated the postural support EMG activity that may occur in a sitting or standing position. Second, this position was more comfortable for the MMD subjects than a complete supine position. Confirmation of positive-pressure changes in gastric pressure during inspiration with tidal breathing verified tube placement.

Abdominal Muscle EMG

Two Teflon-coated (8-μm) fine-wire electrodes were inserted with ultrasound guidance (Acoustic Imaging Ultrasound 5200B, 7.5-MHz transducer) into each of the four abdominal muscles using a modification of a previously published technique (1, 6, 14, 29, 44, 47, 48). The analog EMG signals were filtered to a bandwidth of 100 Hz to 4 kHz, differentially amplified, digitized, and recorded simultaneously (BIOPAC Systems EMG100 amplifiers, MP100 module, and Acknowledge Software version 3.0). EMG visual and acoustic signal quality was evaluated during isometric resistance with trunk flexion, trunk flexion with rotation, and bilateral leg lifts. These maneuvers were also used to obtain maximal recruitment of each of the four abdominal muscles. At the conclusion of each study, maintenance of electrode position was verified by administering a twitch stimulus to each abdominal muscle (STM100A and STMISO, BIOPAC Systems, Goleta, CA). Ultrasound visualization of the twitch contraction within each muscle validated the maintenance of wire electrode position throughout the study.

Specificity of muscle recruitment. With the close approximation of the lateral abdominal wall muscles to each other, there is a concern that cross-contamination of the EMG signal could be a source of error. The use of fine-wire electrodes and appropriate filtering limit this error (1, 42, 50). In addition, we were able to observe independent activity of the muscles during respiration with no contamination of the waveforms from adjacent muscles. At the end of each study, ultrasound visualization of a twitch stimulus within each muscle confirmed that the wires had remained in the correct muscle for the duration of the experiment.

Respiratory Volume

Volume was determined using respiratory inductive plethysmography (Respirtrace System, Ambulatory Monitoring, Ardley, NY) with data collection through a DA100 amplifier (BioPac Systems). The rib cage inductor band was placed around the rib cage with the superior border of the band at the level of the axilla, and the abdominal band was placed below the lower border of the costal margin and above the iliac crest. Bands were then calibrated as previously described (47).
Inspiratory Pressure
Subjects breathed through a Hans Rudolph non-rebreathing valve (Kansas City, MO) that was attached to a mouthpiece and connected to a resistance device (40). Inspiratory pressure was measured using a TSD140 pressure transducer (BioPac Systems) attached to the Hans Rudolph valve and to a DA100 amplifier (BioPac Systems). A nose clip was used to occlude the nostrils.

Data Collection
When subject preparation was completed, each subject breathed 10 breaths at each level of inspiratory resistance (IRB) beginning with no resistance. Weights equal to 20, 40, or 60% of the individual’s MIP (20% MIP, 40% MIP, and 60% MIP, respectively) were added progressively to achieve the desired inspiratory resistance. Once the desired pressure was achieved, data were collected for 10 breaths. Each breath was monitored to ensure that the correct pressure was maintained. During the IRB, the EMG of the abdominal muscles, the gastric pressure, inspiratory pressure, and respiratory inductive plethysmography measures of abdominal and rib cage volumes were recorded simultaneously. A rest period of ~2 min was allowed between each level of resistance. At the end of each IRB trial, oxygen saturation (Nelcor 2000) was recorded and each subject rated the intensity of dyspnea and respiratory effort by pointing to the appropriate value on the modified Borg scale (Table 1) (33, 35). To assess the perception of dyspnea, subjects were asked, “How short of breath are you now?” To assess the perception of respiratory effort, subjects were asked, “How hard are you working to breathe?” To assess the perception of fatigue, subjects were asked, “How tired are you now?”

Data Analysis
Abdominal muscle EMG. The digitized EMG data were rectified, and the area under the EMG curve per unit time (AUC/second) was calculated separately for inspiration and expiration for each muscle. This AUC/second was used as a measure of muscle activity, such that the entire burst of the EMG signal was analyzed (42, 50). For each muscle, the AUC/second of EMG activity was normalized as a percentage of the activity (AUC/second) recorded during a maximal voluntary contraction (14, 42, 47). Because of our simultaneous sampling of 13 different channels of data for this study, the rate at which data was sampled was limited by the maximum throughput of the hardware. Therefore, the EMG signals were acquired at either 200 or 500 Hz. Because these sampling rates are below the Nyquist frequency for fine-wire EMG, we were unable to determine the frequency of the EMG signal. However, these sampling rates do allow a statistical evaluation of the AUC/second of the EMG signal. The validity of the AUC/second as a measure of EMG activity at the lower sampling rates was confirmed in a separate experiment. EMG activity of the brachioradialis was recorded during 10 repetitions of flexion and extension with a fixed weight and fixed distance using sampling rates of 10,000, 2,500, 500, and 200 Hz. ANOVA analysis of AUC/second yielded no statistically significant differences between the mean AUC/second determined at the four sampling rates.

Respiratory volume. Total volume was determined using a sum waveform that was the sum of the abdominal waveform and the rib cage waveform (28, 47). End-expiratory rib cage volume, end-expiratory abdominal volume, and EELV were determined using these waveforms. Illustrations of recordings from a control subject and a subject with MMD during tidal breathing and IRB at 60% MIP are shown in Figs. 1 and 2. The volume at the minimum point of the sum waveform at end expiration during tidal breathing represents functional residual capacity and will be called the end-tidal point. The volume at this end-tidal point has also been called EELV (28) (Fig. 3). The difference between EELV during tidal breathing and the EELV during IRB was calculated. Inspiratory rib cage volume and inspiratory abdominal volume were determined. Inspiratory and expiratory durations were determined from the time between peak and trough points on the sum waveform.

Statistics
Two-way repeated-measures ANOVA was used to determine the overall effect of the IRB and to determine the effect of the disease (MMD vs. control). Dunnett’s multiple-comparisons procedure was used after the ANOVA to compare each activity level with 0% resistance. Data from each series of breaths were averaged to get a mean value for each IRB level. To determine whether variances were homogeneous, a Hartley’s F test was performed. If the variances were not homogeneous, appropriate transforms (either log or arc sine square root) were performed. Pearson correlation coefficients were used to determine the correlation between the EMG activity of the TA and the gastric pressure. Statistical significance for all tests was accepted at P < 0.05.

RESULTS
Pulmonary Function
The MMD subjects demonstrated none to mild restrictive lung disease on the basis of FVC testing (Table 2) (30). All of the MMD subjects demonstrated reduced MIP and MEP, with values comparable to those previously published for MMD populations (7, 32). A plot of vital capacity vs. MIP demonstrates a relationship in normal subjects but no relationship in the MMD group (Fig. 4).

Abdominal Muscle Recruitment
During MEP, both MMD and control subjects recruited the TA and IO at 60–80% of the EMG activity during a maximal contraction (Fig. 5). This activity was significantly greater than the activity of the EO and RA, which were recruited at 10–30% of their maximal EMG activity. There was no significant difference in the pattern of recruitment or the percentage of maximal EMG activity between the MMD group and control group during MEP.

### Table 1. Borg scale

<table>
<thead>
<tr>
<th>Value</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Nothing at all</td>
</tr>
<tr>
<td>0.5</td>
<td>Very, very slight</td>
</tr>
<tr>
<td>1</td>
<td>Very slight</td>
</tr>
<tr>
<td>2</td>
<td>Slight</td>
</tr>
<tr>
<td>3</td>
<td>Moderate</td>
</tr>
<tr>
<td>4</td>
<td>Somewhat severe</td>
</tr>
<tr>
<td>5</td>
<td>Severe</td>
</tr>
<tr>
<td>6</td>
<td>Very severe</td>
</tr>
<tr>
<td>7</td>
<td>Very, very severe</td>
</tr>
<tr>
<td>8</td>
<td>Maximal</td>
</tr>
</tbody>
</table>

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Fig. 1. Sample waveform recordings of control subject showing tidal breathing (0%) and breathing against inspiratory resistance (60% of maximal inspiratory resistance). IRB, inspiratory resistance breathing; Trans, transversus; Vol, volume; Insp, inspiration.

Fig. 2. Sample waveform recordings of myotonic muscular dystrophy (MMD) subject showing tidal breathing (0%) and breathing against inspiratory resistance (60% of maximal inspiratory resistance).
During MIP, all of the abdominal muscles in the control group were recruited at 20–30% of their maximal EMG activity. There was no difference in the level of recruitment among the four abdominal muscles (Fig. 5). In the MMD group, the TA and IO recruited at 55–65% of their maximal EMG activity, which was significantly greater than the activity of the EO and RA (20 and 15% of their maximal EMG activity, respectively). In addition, the TA and IO muscles in the MMD group demonstrated significantly more EMG activity during MIP than the TA and IO muscle of the control group.

During tidal breathing, there was no significant EMG activity in either MMD or control subjects. The EMG activity of the abdominal muscles during IRB (Fig. 6) was much lower (2 and 12% of maximal EMG) than the EMG activity during MIP and MEP (15 and 80%) testing in both MMD and control groups. As the inspiratory resistance challenge increased, the recruitment of the abdominal muscles increased. However, even at 60% MIP, the recruitment of the TA, the most active of the abdominal muscles, was only ~10% of the EMG activity generated during a maximal contraction in both groups.

The EMG activity of the four abdominal muscles of the two groups during IRB is summarized during inspiration (Fig. 6, A and C) and expiration (Fig. 6, B and D). When compared with tidal breathing, the MMD group and control group had significant phasic recruitment of the TA and IO during inspiration and expiration. Neither the control group nor the MMD group demonstrated significant recruitment of the RA. Both the control group and the MMD group significantly recruited the EO at 60% MIP during inspiration only. Because the MIP of the MMD group was approximately one-half of the MIP of the control group, the abdominal muscle recruitment by the MMD group occurred during IRB at a significantly lower absolute pressure (cmH₂O) than the pressure at which the abdominal muscle recruitment of the control group occurred ($P < 0.034$) (Fig. 7).

Qualitative analysis of the timing of the TA and IO muscle activity demonstrated a variety of patterns, as shown in Tables 3 and 4. There was no general pattern of recruitment activity in either the MMD or control subjects. The EMG activity of the abdominal muscles during IRB (Fig. 6) was much lower (2 and 12% of maximal EMG) than the EMG activity during MIP and MEP (15 and 80%) testing in both MMD and control groups. As the inspiratory resistance challenge increased, the recruitment of the abdominal muscles increased. However, even at 60% MIP, the recruitment of the TA, the most active of the abdominal muscles, was only ~10% of the EMG activity generated during a maximal contraction in both groups.

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Qualitative analysis of the timing of the TA and IO muscle activity demonstrated a variety of patterns, as shown in Tables 3 and 4. There was no general pattern of recruitment activity in either the MMD or control group during either inspiration or expiration. Phasic activity occurred in several patterns: phasic activity throughout inspiration and expiration in both the TA and IO, only end-inspiratory activity, or isolated initial expiratory activity. Both groups demonstrated these variations. Interestingly, there was never a pattern of isolated end expiratory activity in either group that would theoretically correspond with reduction in EELV.

Patterns of Rib Cage and Abdominal Motion

During tidal breathing, control subjects exhibited synchronous motion (Fig. 1) with relatively equal contributions of the abdomen and rib cage to inspiratory
tidal volume. The tidal volume increased from 851 ml at 0% IRB to 1,383 ml at 20% IRB and did not significantly change with further increases in IRB levels in the control group (Table 5). With increasing levels of IRB, the inspiratory rib cage and abdominal motion was always outward. However, at 60% MIP, the inspiratory abdominal motion was less outward (reduced volume) than at lower resistance, which is consistent with mild asynchrony. As a result, the contribution of abdomen to the total volume went from 32% at 20% MIP to 17% at 60% MIP. During expiration in the control group, both the rib cage and abdomen decreased in volume and their motion was inward.

The rib cage and abdomen of the MMD group were synchronous during tidal breathing (Fig. 2). During increasing IRB, the inspiratory motion of the abdominal wall was progressively inward with a resulting decrease in abdominal volume. It decreased to such an extent that the contribution of the abdomen to tidal volume was negative at 40 and 60% IRB. The rib cage demonstrated outward motion during inspiration with all levels of IRB (Table 5). In the MMD subjects at 20% IRB, the rib cage contributed 89% of the total volume and 11% was contributed by the abdomen. By 60% MIP, the MMD group exhibited abdominal paradoxical breathing during inspiration with the contribution of the rib cage increasing to 124% of its tidal volume, whereas the abdominal volume decreased to −24% of its tidal volume. During expiration, the abdominal wall had an outward motion (increasing volume), whereas the rib cage was moving inward (decreasing volume).

There was a significantly greater reduction in EELV of the control group with increased IRB than in the MMD subjects. There was a slight reduction in EELV in the MMD group (Fig. 8) at 20% IRB but none at 40% or 60% IRB.

Gastric Pressure

There was no effect of IRB on end-inspiratory gastric pressure in either group. During expiration, gastric pressure decreased to its nadir at end expiration. There was no increase in gastric pressure with abdominal EMG activity using group analysis. However, the EMG levels were low and data were variable, with some individuals demonstrating increased gastric pressure. Because of the variability in expiratory gastric pressure, no correlation was found between gastric pressure, abdominal EMG activity, and abdominal motion in either group. With MIP and MEP determinations, where the EMG activity was high, we did observe significantly increased gastric pressure. During MIP, the gastric pressure of both the MMD and control groups was correlated to the EMG activity of the TA (MMD $r = 0.73$, control $r = 0.92$). This was not true during MEP, where the EMG activity of the TA corre-
lated with the gastric pressure \((r = 0.62)\) for the control group, but the EMG and gastric pressure had no correlation in the MMD group \((r = -0.01)\).

**Ventilatory Parameters**

The tidal volume of the control group was two to three times the volume seen in the MMD group at each level of resistance breathing (Table 5). The tidal volume of both groups increased significantly from 0% to 20% MIP, but it did not significantly change as the inspiratory resistance was increased to 40–60% MIP. As has been previously observed, the use of a mouthpiece caused a significant increase in the tidal volume in the control group \((445 \pm 46\) ml with no mouthpiece to \(916 \pm 116\) ml when a mouthpiece was used) \((4, 23, 49)\). However, the MMD group had no change in tidal volume with the insertion of the mouthpiece; tidal volume without a mouthpiece was \(343 \pm 42\) ml and with a mouthpiece the volume was \(369 \pm 49\) ml.

Respiratory rate was significantly greater in the MMD group than the control group at all levels of IRB. Increasing the resistance did not significantly alter the respiratory rate in either the control or MMD groups. The higher respiratory rate in the MMD group did not compensate for the reduction in tidal volume, and, as a result, the minute ventilation of the MMD group was significantly less than that of the control group at all levels of resistance. Both groups had a significant increase in minute ventilation with increasing inspiratory resistance. Despite the fact that the minute ventilation of the MMD group was \(~50\)% of the control group, oxygen saturation was not significantly different between the two groups.

**Perceptions of breathing.** MMD and control groups demonstrated statistically significant increases in all three perceptions of breathing, particularly at 40 and 60\% MIP. The two groups did not differ in their perception of dyspnea or fatigue, but the MMD group perceived significantly greater respiratory effort at the higher IRB levels than the control group (Table 6).

**DISCUSSION**

The purpose of this study was to determine how respiratory muscular weakness associated with MMD affects abdominal muscle recruitment, chest wall motion, and pulmonary function. This was done by subjecting MMD and control subjects to static pulmonary pressure measurements and an exercise stress that

<table>
<thead>
<tr>
<th>Table 3. EMG activity of the transversus abdominis</th>
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<tr>
<td><img src="https://via.placeholder.com/150" alt="Graph" /></td>
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</tbody>
</table>

**Fig. 7.** EMG activity of the TA as a function of the mouth pressure during IRB. Max, maximum.
was created by breathing against increasing levels of inspiratory resistance.

Our MMD subjects demonstrated significant respiratory muscle weakness, as indicated by reduced MIP and MEP, although they had normal to minimally reduced FVC. These results are consistent with other reports of static pressure measurements in MMD populations (7, 9, 10, 53).

Table 4. EMG activity of the internal oblique

<table>
<thead>
<tr>
<th>Subject No.</th>
<th>0% IRB</th>
<th>60% IRB</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>Inspiration</td>
<td>Expiration</td>
</tr>
<tr>
<td></td>
<td>Beginning</td>
<td>Middle</td>
</tr>
<tr>
<td>1</td>
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<td>9</td>
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<td>-</td>
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<tr>
<td>10</td>
<td>-</td>
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</tbody>
</table>

-, Absence of EMG activity; +, presence of EMG activity.

Table 5. Ventilation and gastric pressure

<table>
<thead>
<tr>
<th>Resistance, %MIP</th>
<th>0</th>
<th>20</th>
<th>40</th>
<th>60</th>
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</thead>
<tbody>
<tr>
<td>Mouth pressure, cmH2O</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*†‡</td>
<td>2.9 ± 0.3</td>
<td>21.8 ± 3.2</td>
<td>42.3 ± 6.6</td>
<td>61.0 ± 11.4</td>
</tr>
<tr>
<td>MMD</td>
<td>2.0 ± 0.3</td>
<td>15.1 ± 1.5</td>
<td>27.8 ± 3.6</td>
<td>33.5 ± 3.6</td>
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<tr>
<td>Tidal volume, ml</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*†</td>
<td>851 ± 108</td>
<td>1,383 ± 229</td>
<td>1,388 ± 203</td>
<td>1,067 ± 157</td>
</tr>
<tr>
<td>MMD</td>
<td>369 ± 49</td>
<td>576 ± 117</td>
<td>572 ± 96</td>
<td>540 ± 78</td>
</tr>
<tr>
<td>Rib volume, ml</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*†</td>
<td>439 ± 96</td>
<td>927 ± 146</td>
<td>979 ± 119</td>
<td>879 ± 126</td>
</tr>
<tr>
<td>MMD</td>
<td>182 ± 20</td>
<td>511 ± 99</td>
<td>646 ± 97</td>
<td>668 ± 119</td>
</tr>
<tr>
<td>Abdominal volume, ml</td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td>Control*†</td>
<td>412 ± 40</td>
<td>456 ± 105</td>
<td>409 ± 108</td>
<td>189 ± 38</td>
</tr>
<tr>
<td>MMD</td>
<td>186 ± 39</td>
<td>64 ± 59</td>
<td>-73 ± 66</td>
<td>-129 ± 90</td>
</tr>
<tr>
<td>Respiratory rate, breaths/min</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*</td>
<td>10.7 ± 1.0</td>
<td>10.2 ± 1.4</td>
<td>10.8 ± 1.4</td>
<td>11.9 ± 1.3</td>
</tr>
<tr>
<td>MMD</td>
<td>16.1 ± 1.6</td>
<td>14.7 ± 1.4</td>
<td>15.8 ± 1.7</td>
<td>15.1 ± 1.6</td>
</tr>
<tr>
<td>Minute ventilation, l/min</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*†</td>
<td>9.7 ± 1.4</td>
<td>14.4 ± 2.4</td>
<td>16.3 ± 3.1</td>
<td>15.4 ± 3.3</td>
</tr>
<tr>
<td>MMD</td>
<td>5.5 ± 0.4</td>
<td>7.8 ± 1.3</td>
<td>8.4 ± 1.1</td>
<td>7.3 ± 1.0</td>
</tr>
<tr>
<td>Gastric pressure (end inspiratory), cmH2O</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control*</td>
<td>7.6 ± 0.7</td>
<td>7.8 ± 1.3</td>
<td>8.4 ± 1.2</td>
<td>6.9 ± 1.2</td>
</tr>
<tr>
<td>MMD</td>
<td>10.6 ± 1.2</td>
<td>10.8 ± 1.4</td>
<td>13.9 ± 2.6</td>
<td>16.5 ± 4.6</td>
</tr>
<tr>
<td>Oxygen saturation, %</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Control</td>
<td>98.9 ± 0.4</td>
<td>99.0 ± 0.9</td>
<td>99.5 ± 0.3</td>
<td>98.0 ± 0.9</td>
</tr>
<tr>
<td>MMD</td>
<td>96.7 ± 1.1</td>
<td>97.6 ± 0.9</td>
<td>97.4 ± 0.9</td>
<td>97.6 ± 1.0</td>
</tr>
</tbody>
</table>

Values are means ± SE. Results of 2-way ANOVA: *Disease effect, P < 0.05. †Inspiratory resistance effect, P < 0.05. ‡Disease-resistance interaction, P < 0.05.
Activated During Respiration

Hypothesis 1: Abdominal Muscles Are Selectively Activated During Respiration

In the past, abdominal muscles were considered to act as a single unit during respiration (24). With the development of more sophisticated EMG techniques, it has been shown that individual abdominal muscles contribute to breathing in normal controls (1, 2, 12, 16, 29, 45, 47, 48) and in subjects with chronic obstructive pulmonary disease (41). To our knowledge, this is the first study to describe abdominal muscle activity in MMD subjects. This study demonstrates selective activity of the abdominal muscles in MMD subjects and normal subjects during static pressure testing.

MMD and control subjects had different patterns of abdominal muscle recruitment during MIP testing; in the control group, all four abdominal muscles were recruited as a unit with equal activity, whereas the TA and IO muscles of the MMD group were recruited to a greater degree than the EO and RA. This pattern suggests that the TA and IO are accessory muscles of inspiration in persons with weakened respiratory muscles. The abdominal muscle activity observed during inspiratory effort, in this case, may be due to an attempt to maximize the contraction of the diaphragm in the zone of apposition (18, 36).

In contrast to MIP, there were no differences between the MMD and control groups in the recruitment patterns and percentage of EMG activity of the abdominal muscles during MEP testing. In both groups, the TA and IO muscles had greater activity than the EO and RA muscles. There are no prior reports of recruitment patterns during MEP testing. However, De Troyer et al. (16) reported that, in seated, normal subjects, the TA, EO and RA activity was equal during expulsive maneuvers from functional residual capacity. Hodges et al. (29) and Goldman et al. (24) found that all four abdominal muscles were collectively active in expulsive maneuvers or during cough. These differences in recruitment may be due to the differences in the method or differences in posture.

The expiratory activity of the abdominal muscles observed during IRB exercise in both groups during expiration was as expected from results of previous studies in normal subjects (1, 2, 16, 25, 38, 48). In our study, the patterns of recruitment were the same for both groups. The TA and IO were recruited at lower levels of IRB than the EO and RA. The EO was activated only at the highest level, and the RA was never recruited. The consecutive recruitment of the TA, IO, EO, and the RA has been seen in normal subjects (1, 2, 48) and in chronic obstructive pulmonary disease subjects (16) during carbon dioxide rebreathing.

We also observed EMG activity of the abdominal muscles during inspiration. Abe et al. (2) reported inspiratory abdominal muscle activity during carbon dioxide rebreathing in normal subjects. However, the activity they observed occurred primarily at the beginning of inspiration and was called postexpiratory expiratory activity (2). Hodges et al. (29), using inspiratory resistance, reported midinspiratory activity when normal subjects were at rest in a standing position. During upright posture, gravitational forces pull the abdominal contents down and outward along with the diaphragm. Abdominal muscles must contract to maintain a favorable diaphragm position. With abdominal contraction in the upright position, abdominal compliance decreases. Thus both tonic activity to counteract gravitational forces and selective contraction with respiratory activity occur in upright postures (1, 2, 16, 29). The inspiratory EMG activity observed was quite variable from person to person, with activity occurring during the beginning, middle, and end of inspiration (Table 3). Differences between our study and others may be due to differences in the challenged breathing protocol and/or differences in posture (supine vs. standing or sitting).

Inspiratory abdominal muscle contraction may theoretically improve biomechanical advantage by increasing abdominal pressure along the costal margins to provide a counterforce for the descending diaphragm at the area of apposition. This counterforce allows expansion of the distal rib cage, thereby placing the costal diaphragm fibers in the zone of apposition in a favorable biomechanical position to enhance the inspiratory force of the crural diaphragm (17). Other theories as to the presence of inspiratory abdominal activity include possible “braking” of inspiration and prevention of hyperinflation of the lungs (1, 39).

Table 6. Respiratory sensations using Borg scale

<table>
<thead>
<tr>
<th></th>
<th>Resistance, %MIP</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>0</td>
</tr>
<tr>
<td>Dyspnea Control*</td>
<td>0 ± 0.1</td>
</tr>
<tr>
<td>MMD</td>
<td>0 ± 0.1</td>
</tr>
<tr>
<td>Work of breathing</td>
<td>0 ± 0.1</td>
</tr>
<tr>
<td>Control† MMD</td>
<td>0 ± 0.1</td>
</tr>
<tr>
<td>Fatigue Control*</td>
<td>0 ± 0.1</td>
</tr>
<tr>
<td>MMD</td>
<td>0 ± 0.1</td>
</tr>
</tbody>
</table>

Values are means ± SE. Results of 2-way ANOVA: *Inspiratory resistance effect, P < 0.05. †Disease-resistance interaction, P < 0.05.
In this study, no significant EMG activity was recorded during tidal breathing in the supine position in either group. The absence of abdominal muscle activity in the supine posture with quiet breathing concurs with other reports of tidal breathing in normal subjects (12, 21, 24, 38, 45). However, Abe et al. (1) observed very slight, but significant, expiratory activity during supine tidal breathing in normal subjects. The difference between our study and that of Abe et al. may be interpretation of what constitutes significant activity.

**Hypothesis 2: MMD Subjects Recruit Abdominal Muscles at an Inspiratory Resistance That is Significantly Less Than the Resistance Required for the Recruitment of Abdominal Muscles in Control Subjects**

Although the abdominal muscle activities of the MMD and control groups appear to be similar when measured as a percentage of their MIP (Fig. 6), the absolute pressure at which the muscles of the MMD group were recruited is one-half that at which the controls recruited the muscles (Fig. 7). When measured at the same inspiratory pressure (cmH$_2$O), MMD subjects demonstrated approximately twice the EMG activity of control subjects (Fig. 6). The greater EMG activity for the same level of resistance in the weakened muscles of MMD subjects is consistent with sympathetically recruited principles (20, 34). To lift an identical load, a dystrophic muscle typically recruits a greater number of motor units than a normal muscle.

**Hypothesis 3: Mechanical Responses to Expiratory Abdominal EMG Activity Are an Increase in Gastric Pressure and a Relative Reduction in Abdominal Volume**

In the past, studies have assumed that an increase in expiratory gastric pressure indicated an increase in abdominal muscle recruitment (37). Prior studies have found an association between abdominal muscle activity and gastric pressure during expiration with subjects in upright positions (14, 38). In our IRB study, which was performed with subjects in a supine position, neither the MMD group nor the control group demonstrated an association between EMG activity and peak gastric pressure during the IRB protocol. Although both the control and MMD groups had statistically significant EMG activity of the TA and IO during IRB exercise, the level of EMG activity was low (2–14% of maximal EMG activity), even at the highest level of IRB exercise (60% IRB). This low level of abdominal EMG activity during expiration in IRB may not have been great enough to produce an increase in gastric pressure. However, during MIP, where there was high EMG activity, there was a high correlation between TA EMG activity and gastric pressure in the control group ($r = 0.92$) and in the MMD group ($r = 0.79$), respectively. During MEP, even though the EMG activity of both the MMD and control groups was high, there was a correlation between gastric pressure and TA activity only in the control group ($r = 0.62$) and not in the MMD group ($r = -0.01$). However, the lack of correlation of abdominal activity in MEP with gastric pressure in only the MMD group is difficult to explain, but it may be related to several observations. 1) At 60% IRB in the MMD group, the abdominal motion was outward during expiration, whereas the abdominal motion of the controls was inward during expiration. The outward motion of the abdomen during expiration in the MMD group may be enough to dissipate any increase in gastric pressure generated by abdominal muscle contraction. It is likely that, at maximal expiratory effort, a similar abdominal motion pattern would be demonstrated. Unfortunately, we do not have reliable abdominal motion measurements during MEP testing to confirm this explanation. 2) A decrease in lung compliance during expiration in the MMD group may also lead to lower gastric pressure given the supine posture. However, we did not have a measure of lung compliance. 3) The marked weakness of abdominal muscles and an inability to generate significant gastric pressure in the MMD group is another possible explanation. However, the correlation of abdominal EMG activity and gastric pressure during MIP measurements makes this explanation less likely. It is clear that further study is needed to understand the relationship between abdominal muscle activity and gastric pressure in MMD subjects.

**Hypothesis 4: Expiratory Activity of the Abdominal Muscles is Associated With a Reduction in EELV**

A theoretically advantageous mechanical consequence of expiratory abdominal muscle activity is a reduction in EELV (Fig. 3). Reduction in EELV could assist inspiration by two mechanisms. 1) Prolonged expiration and increased expiratory flow pushes the diaphragm into the thorax. This cephalad displacement of the diaphragm during expiration increases the elastic energy of the diaphragm. Initial inspiration can then be passive. Thus the abdominal muscles indirectly assist in the work of inspiration. 2) Reduction in EELV also lengthens the diaphragmatic fibers, placing them in a more optimal position in the muscle's length-tension curve with contraction, especially if the diaphragm is flattened (1, 16, 28, 51, 52). We observed decreased EELV in control subjects, but not in the MMD group. Our control subjects had a reduction in the EELV of 0.25 liters, which is 5% of vital capacity (0.25 liters/4.8 liters), whereas the MMD subjects had a reduction of 2% (0.1 liters/3.8 liters). The lack of reduction of EELV in MMD subjects is likely due to disordered breathing patterns. Even though the expiratory abdominal muscle activity was small in control subjects, it may be responsible for the reduction in EELV that we observed. Henke et al. (28) found a reduction in EELV during exercise and hypothesized that this reduction aids inspiration by optimizing diaphragmatic length and permitting elastic recoil of the chest wall. The clinical significance of the reduced volume observed in our study is unclear.
Hypothesis 5. MMD Subjects Exhibit Dysynchronous Breathing Patterns When Challenged With Low Levels of Inspiratory Resistance and at Higher Levels of Inspiratory Resistance Demonstrate Abdominal Paradox

In our study, MMD subjects demonstrated synchronous breathing patterns during tidal breathing. These results differ from those of Serisier et al. (43), who in his study of MMD patients at rest, observed abdominal paradox in 1 of 19 subjects and chaotic breathing patterns in 7 of 19 subjects. The differences may be due to greater severity of the dystrophy in the MMD subjects in the study of Serisier et al. Nevertheless, we did show that MMD subjects exhibited abdominal asynchrony when breathing against low levels of inspiratory resistance, and 6 of 10 exhibited paradoxical abdominal motion at the highest level of resistance (60% IRB). In comparison, control subjects exhibited a synchronous pattern of breathing until 60% MIP, when their breathing became asynchronous with 1 of 10 showing paradoxical abdominal motion at the highest level of resistance.

Ewig et al. (22) reported that individuals who were lacking abdominal muscles (prune belly syndrome) breathed synchronously when in the supine position but exhibited abdominal paradox during exercise. Our MMD subjects with abdominal muscle weakness demonstrate chest wall mechanics similar to those of subjects with absent abdominal muscles.

Ewig et al (22) also noted that the asynchrony caused inefficient breathing patterns, respiratory fatigue, and increased work of breathing. Our observations showed a significant increase in the perception of work of breathing in both groups with increasing levels of IRB. However, when the MMD group was breathing more asynchronously (Table 4), they had a greater increase in the perception in the work of breathing than the control group. This increased perception may be related to the increased activation of the weakened muscles or to the alterations of chest wall motion of MMD group. Although the perception of work of breathing was greater in the MMD group, we found that the two groups had similar increases in the perception of dyspnea and fatigue with increasing IRB levels.

Work by Cala et al. (11) suggests that dysynchronous patterns of breathing may also be related to chest wall compliance, as well as to abdominal muscle weakness. Particularly in the supine position, changes in rib cage motion can affect abdominal wall motion without any abdominal activity because of the connection between the rib cage and abdomen (11). It is possible that the rib cage muscles, which are involved in active expiration, could be partially responsible for the asynchrony. A limitation of the present study is the lack of a direct measure of diaphragmatic or rib cage muscle activity to assist in the understanding of changes in rib cage volume and compliance. However, in a study of rib cage muscles using surface EMG, MMD subjects demonstrated expiratory lower rib cage muscle activity, whereas there was no activity in the control group (31).

The reduced rib cage volumes that we observed at all IRB levels in comparing the MMD subjects with control subjects may be associated with decreased rib cage compliance. However, comparing the relative contribution of the ribs vs. the abdominal volumes to the tidal volume, we found marked increases in the rib cage contribution during increasing workloads. Our findings indicate that the rib cage expands to a greater degree than the abdomen with increasing IRB. Thus the rib cage compliance is probably not reduced in our subjects, although we did not have a direct measure of compliance. Teramoto et al. (46) found a decrease in rib cage contribution in upright exercise in the aged compared with younger controls. Unlike the aged controls, our MMD subjects can expand the rib cage to increase tidal volume during exercise. Although controls demonstrated a high correlation between vital capacity and MIP ($r = 0.76$; Fig. 4), there was no significant correlation in the MMD group in our study. De Troyer et al. (17) suggest that, when MIP does not directly correlate with vital capacity, factors other than muscle weakness are the cause; e.g., poor lung compliance may contribute to alterations in vital capacity in the MMD group. Esophageal pressure measurements would have been helpful in understanding lung compliance in our subjects (18). A potential bias in our study was the difference in gender distribution between the two study groups, with the control group having more female subjects than the MMD group. In examining the control group for this bias, we found no gender differences in their EMG responses. However, the female controls had significantly smaller rib cage volume and abdominal volume than the male controls. Because the MMD subjects had significantly smaller volumes than the control group did, this gender bias would reduce the significance found between the MMD and control groups and therefore did not affect the interpretation of our results.

Implication and Summary

Observations from this study increase our understanding of basic respiratory physiology in MMD. In a group of MMD subjects with normal or mild reduction in FVC, MEP was markedly reduced, indicating abdominal muscle weakness. The recruitment patterns of the abdominal muscles overall were similar to those of normal subjects, demonstrating selective activity of the TA and IO. However, MMD subjects recruited the abdominal muscles at lower absolute pressure during IRB compared with control subjects. In addition, abdominal muscle EMG activity of MMD subjects is significantly higher than that of controls during IRB when compared at the same absolute pressures. This is consistent with recruitment patterns observed with myopathic muscles. Despite this greater EMG activity of the abdominal muscles seen in the MMD subjects, corresponding mechanical changes (increased gastric pressure and inward abdominal motion) were not seen in the MMD group; this was likely due to the severity of the abdominal weakness. Abdominal wall motion was dysynchronous and paradoxical in the MMD
group with exercise. The theoretical advantage of expiratory abdominal activity in decreasing EELV was seen only in controls and not in MMD subjects. The inspiratory EMG activity we observed may provide a biomechanical advantage by providing support to enhance the inspiratory descent of the crural diaphragm. During MIP testing, the MMD group demonstrated greater activity compared with controls, indicating the TA and IO are likely accessory muscles of inspiration in MMD. Marked differences in chest wall mechanics and minute ventilation during exercise, most likely due to abdominal muscle weakness, can be seen in MMD subjects with minimal to no loss in FVC. As a result, inefficient patterns of breathing lead to increased perception in work of breathing.

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