Diaphragmatic function in advanced Duchenne muscular dystrophy

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Abstract

The aim of this study was to assess diaphragm electrical activation and diaphragm strength in patients with advanced Duchenne muscular dystrophy during resting conditions. Eight patients with advanced Duchenne muscular dystrophy (age of 25 ± 2 years) were studied during tidal breathing, maximal inspiratory capacity, maximal sniff inhalations, and magnetic stimulation of the phrenic nerves. Six patients were prescribed home mechanical ventilation (five non-invasive and one tracheotomy). Transdiaphragmatic pressure and diaphragm electrical activation were measured using an esophageal catheter. During tidal breathing (tidal volume 198 ± 83 ml, breathing frequency 25 ± 7), inspiratory diaphragm electrical activation was clearly detectable in seven out of eight patients and was 12 ± 7 times above the noise level, and represented 45 ± 19% of the maximum diaphragm electrical activation. Mean inspiratory transdiaphragmatic pressure during tidal breathing was 1.5 ± 1.2 cmH2O, and during maximal sniff was 7.6 ± 3.6 cmH2O. Twitch transdiaphragmatic pressure deflections could not be detected. This study shows that despite near complete loss of diaphragm strength in advanced Duchenne muscular dystrophy, diaphragm electrical activation measured with an esophageal electrode array remains clearly detectable in all but one patient.

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Keywords: Diaphragm; Duchenne muscular dystrophy; Diaphragm electrical activity; Magnetic stimulation

1. Introduction

In Duchenne muscular dystrophy (DMD), respiratory muscle weakness starts as early as the second decade of life [1,2], and is followed by a progressive loss in maximal inspiratory pressures and vital capacity [3–6]. Although there is little information about diaphragmatic involvement in humans, studies in animal models of DMD show that the diaphragmatic strength (in vitro) is reduced to 13.5% of control in the very late stages of the disease [7]. In the very late stages of DMD, human patients develop chronic respiratory insufficiency with hypercapnia, which is often treated with non-invasive positive pressure ventilation [8].

It has been suggested that the chronic hypoventilation in advanced DMD may be a direct result of the respiratory muscle weakness, but it could also be due to a reduced respiratory drive [9]. There is very little information about neural drive to breathe in advanced DMD. When faced with an instantaneous elastic or resistive load, DMD patients (mean age 19 years) defend their tidal volume (VT), similar to healthy subjects [10]. In other groups of patients with functional diaphragm weakness (such as chronic obstructive pulmonary disease and post-polio syndrome), defending VT during resting conditions demands a three to five-fold increase in diaphragm electrical activity (EA di) [11,12] compared to healthy subjects. Therefore, one could expect that patients with advanced DMD—who have respiratory muscle weakness—have an elevated level of diaphragm electrical activation during resting breathing. On the other
hand, DMD patients do not choose to increase their ventilation (i.e. they hypoventilate), as demonstrated by their hypercapnia.

The aim of the present study, therefore, was to quantify the amount of diaphragm electrical activation and transdiaphragmatic pressure swings during resting breathing in patients with advanced DMD. A second aim was to assess diaphragm strength with a non-volitional method, that of magnetic stimulation of the phrenic nerves.

2. Patients and methods

The ethical and scientific committees of Göteborg University, Göteborg, Sweden approved the protocol. Written informed consent was obtained from the patients.

2.1. Patients

Eight patients with Duchenne muscular dystrophy, with a mean age of 25 ± 2 years were studied. Clinical data for the patients are presented in Table 1. The patients all had sparse motor function in their extremities, were confined to their wheelchairs and were dependent on attending personnel for their daily-activities. Six of the eight patients were prescribed home mechanical ventilation, five non-invasively with nasal mask and one with tracheotomy. None of the patients had surgery to correct their scoliosis. The mean vital capacity for six of the eight patients (measured within 1 year of the study date) was 467 ± 301 ml.

2.2. Measurements

The electrical activity of the diaphragm (EAdi) was obtained via a multiple array gastro-esophageal electrode consisting of eight sequential differential bipolar electrode pairs, mounted on silicone tubing, as previously described in [11].

Two latex balloons were mounted on the catheter, below and above the electrode array, to allow for measurements of gastric (Pga) and esophageal pressures (Pes), respectively. Diaphragm strength was assessed by measurements of transdiaphragmatic pressure (Pdi), calculated as difference between Pga and Pes. The two balloon catheters were connected to two differential pressure transducers (± 350 cmH2O, Omega PX72-005GV, Stamford, CT, USA).

Mouth pressure (Pmo) was measured via a side port in the mouthpiece and connected to a third differential transducer (± 350 cmH2O, Omega PX72-005GV, Stamford, CT, USA). Inspiratory flow was measured with a pneumotachograph (Jaeger Screenmate, Jaeger GmbH & Co. KG, (resistance was 0.36 cmH2O/L s⁻¹, deadspace 50 ml). The deadspace of the mouthpiece was 10 ml. In the patient with the tracheotomy, the tracheotomy was closed and the patient breathed through a mouthpiece.

EAdi signals were processed as previously described [12], and stored on hard disk for off-line analysis. Flow, Pmo, Pes, Pga was acquired simultaneously with the EAdi data (Data Translation, DT 2811-PGH, Marlboro, MA, USA).

2.3. Experimental protocol

The esophageal catheter with the electrodes and pressure balloons were passed transnasally and positioned at the level of the gastro-esophageal junction with feedback from an on-line display of the EAdi signals from all electrode pairs on the computer monitor. With this position of the catheter, the esophageal balloon is located in the lower third of the esophagus and the gastric balloon in the stomach.

Subjects were seated in their wheelchair and instructed to breathe quietly for 20 min after insertion of the catheter before the phrenic nerve stimulation to avoid twitch potentiation [13]. Phrenic nerve stimulation was performed using bilateral anterior magnetic stimulation (BAMPS) using two magnetic stimulators Magstim 200 (Magstim Company, Whittland, Dyfed, Wales) to power each of two 43 mm double circular coils. Stimulation was applied at resting end expiration against a closed airway. The subjects wore a nose clip during stimulations. The phrenic nerves were stimulated at the posterior border of the sternocleidomastoid muscle at the level of the cricoid cartilage. The stimulators were charged to a range of predetermined percentages of maximal output (70, 80, 90, and 100%).

Table 1

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yrs)</th>
<th>Years of NIMV (h/day)</th>
<th>PCO₂ (mmHg)</th>
<th>PO₂ (mmHg)</th>
<th>HCO₃ (mEq/L)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>24</td>
<td>5/18</td>
<td>51.8</td>
<td>87.0</td>
<td>27.7</td>
</tr>
<tr>
<td>2</td>
<td>29</td>
<td>5/18</td>
<td>48.0</td>
<td>80.3</td>
<td>25.3</td>
</tr>
<tr>
<td>3</td>
<td>25</td>
<td>0.5/Not available</td>
<td>48.8</td>
<td>85.5</td>
<td>27.7</td>
</tr>
<tr>
<td>4</td>
<td>28</td>
<td>0.5/8</td>
<td>60.0</td>
<td>79.5</td>
<td>27.1</td>
</tr>
<tr>
<td>5</td>
<td>23</td>
<td>4/10</td>
<td>59.3</td>
<td>82.5</td>
<td>28.0</td>
</tr>
<tr>
<td>6</td>
<td>25</td>
<td>0/0</td>
<td>51.8</td>
<td>78.8</td>
<td>26.2</td>
</tr>
<tr>
<td>7</td>
<td>23</td>
<td>7/23</td>
<td>50.3</td>
<td>77.3</td>
<td>24.3</td>
</tr>
<tr>
<td>8</td>
<td>25</td>
<td>0/0</td>
<td>45.8</td>
<td>102.0</td>
<td>24.6</td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>25 ± 2</td>
<td></td>
<td>51.9 ± 5.1</td>
<td>84.1 ± 8.0</td>
<td>26.4 ± 1.5</td>
</tr>
</tbody>
</table>

NIMV, non-invasive mechanical ventilation; SD, standard deviation.
At least three stimulations were made at 90 and 100%. Supra-maximal stimulation was achieved in all subjects as judged by the $E_{Adi}$. At least five baseline unpotentiated supra-maximal phrenic nerve stimulations were then obtained in each subject (100%).

Following the phrenic nerve stimulations, while seated in the wheelchair, each patient performed a maximal sniff inhalation maneuver from functional residual capacity (FRC) with one nostril occluded, and a slow inspiration to total lung capacity (TLC). These maneuvers were used to determine peak $P_{di}$ [14] and maximum voluntary diaphragm activation [11], respectively. The maneuvers were repeated until three reproducible values were obtained or the patient would not continue to perform the maneuvers. More than 30 s of rest was allowed between attempts for a given maneuver and 10–15 min between the two types of maneuvers.

After these maneuvers, patients breathed quietly for 10 min, through a mouthpiece with a noseclip on. Measurements were continuously made during this period.

2.4. $E_{Adi}$ signal processing for spontaneous breathing

Automatic on-line digital processing of $E_{Adi}$ was performed as previously described [11,12] with algorithms that account for the negative influences of muscle-to-electrode distance filtering [15,16] and contamination by the ECG. The root-mean-square (RMS) for the processed $E_{Adi}$ signal was calculated every 16 ms and used to quantify signal strength. Signal segments with residual disturbances due to cardiac electric activity or common mode signals were evaluated via specific detectors, and replaced by a predicted value, e.g. the previously accepted value [11].

2.5. Evaluation of signal quality

2.5.1. $E_{Adi}$ amplitude in the time domain

The baseline noise level of the $E_{Adi}$ signal was quantified by determining the difference between the minimum and the most frequently occurring value for a 20-point segment preceding the onset of each inspiratory $E_{Adi}$. This 20-point segment preceding the inspiration (equivalent to 320 ms) was assumed to be expiratory. For quiet breathing, the peak inspiratory $E_{Adi}$ was expressed as an increase above the background noise. As well, the quiet breathing values of $E_{Adi}$ were expressed as a percent of the maximum observed $E_{Adi}$ during the TLC or sniff maneuvers, in order to obtain a value of relative diaphragm activation [11].

Note that the ‘detectable change’ in signal amplitude above the noise level (as described above) is not to be confused with the signal quality indices developed for analysis of $E_{Adi}$ signals in the frequency domain (see below).

2.5.2. $E_{Adi}$ signal quality analysis in the frequency domain

In order to evaluate if the signals from the DMD patients were suitable for accurate interpretation of power spectral analysis and center frequency analysis, the processed $E_{Adi}$ signals were evaluated using four signal quality indices as previously described by Sinderby et al. [17].

2.6. Analysis of respiratory parameters during spontaneous breathing

Mean $P_{di}$ and $P_{ex}$ swings were calculated between the onset of $E_{Adi}$ and the peak of inspiratory $E_{Adi}$ (visually determined). Neural timing parameters (neural inspiratory time $N_{Ti}$, neural expiratory time $N_{Te}$) were determined from the $E_{Adi}$ signal. Volume was obtained by linearly integrating flow. Inspiratory and expiratory times were also calculated based on zero crossings of the flow signal.

2.7. Analysis of diaphragm compound muscle action potentials (CMAPs) and twitch $P_{di}$

CMAPs were measured with each of the eight electrode pairs on the esophageal catheter. The latency was defined as the time period between the onset of the stimulation artifact and the onset of the CMAP. Latency time was calculated from the CMAP signal obtained from a given single electrode pair showing a detectable CMAP.

We measured the twitch $P_{di}$ as the deflection in $P_{di}$ following stimulation.

3. Results

Data in text, tables and figures are presented as means ± standard deviation (SD). In one patient (patient no.1) we were not able to obtain a maximal inspiratory maneuver because the patient refused. In another patient (patient no. 8), the $E_{Adi}$ signals during quiet breathing were not detectable.

3.1. Spontaneous breathing data

During resting breathing, tidal volume and neural breathing frequency were $198±83$ ml and 25 ± 7 breaths per minute, respectively. Respiratory rate calculated from the flow tracings was 27 ± 12). In the seven patients who were willing to perform the maneuver, inspiratory capacity was found to be $413±160$ ml (Table 2).

For the seven patients with measureable diaphragm electrical activity, inspiratory $E_{Adi}$ during resting breathing was clearly detectable and was $12±7$ times above the random fluctuations of the background noise level, as demonstrated in an individual subject in Fig. 1. Neural inspiratory and expiratory times were $720±130$ and $1670±510$ ms, respectively, and their ratio ($T_i/T_{tot}$) was $0.31±0.05$. Flow $T_i$ and flow $T_e$ were $1007±273$ and $1740±819$, respectively, with a corresponding $T_i/T_{tot}$ of $0.43±0.12$. The inspiratory $E_{Adi}$ represented $45±19\%$ of the maximum $E_{Adi}$ (Table 3).
Calculated from the EAdi power spectrum, the signal to noise (SN) ratio was below the inclusion level recommended by Sinderby et al. [19] for accurate interpretation of the center frequency value, and was on average 11.5 ± 4.0 dB for the group.

Different from EAdi, the Pes signal was significantly disturbed by cardiac artifacts in all patients, as well as frequent esophageal spasm-induced artifacts (Fig. 1). Swings in Pdi during resting breathing were barely detectable (Table 3 and Fig. 1) and averaged 1.5 ± 1.2 cmH2O. Negative deflections in Pga were observed on inspiration in six out of the patients. Maximal sniff Pdis were 7.6 ± 3.6 cmH2O for the group.

### 3.2. Electromagnetic stimulation

Twitch Pdi deflections could not be detected in any of the patients, despite the presence of the diaphragm compound muscle action potentials. Twitch latency time was 8.2 ± 1.3 ms for the group. Fig. 2 shows data from one patient with advanced DMD and no detectable twitch Pdi.

### 4. Discussion

In the present study, we evaluated the functional performance of the diaphragm in advanced Duchenne muscular dystrophy. We found detectable signals during resting breathing in seven out of the eight patients studied. These patients used 45% of the maximal voluntary activation of the diaphragm to maintain a tidal volume of about 200 ml. This was associated with a swing in Pdi of 1.5 cmH2O, equivalent to 18% of the maximum pressure generating capacity of the diaphragm. Although the diaphragm compound muscle action potential was present, the Pdi generated by supra-maximal stimulation was not detectable.

We observed clear detectable EAdi signals during quiet breathing in all but one of the patients. In patient no. 8, EAdi during quiet breathing was not detectable. This was not due to poor electrode positioning because we observed CMAPs

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**Table 2**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Inspiratory capacity (ml)</th>
<th>Sniff Pdi (cmH2O)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>–</td>
<td>Not obtainable</td>
</tr>
<tr>
<td>2</td>
<td>320</td>
<td>4</td>
</tr>
<tr>
<td>3</td>
<td>420</td>
<td>10</td>
</tr>
<tr>
<td>4</td>
<td>540</td>
<td>11</td>
</tr>
<tr>
<td>5</td>
<td>300</td>
<td>8</td>
</tr>
<tr>
<td>6</td>
<td>560</td>
<td>5</td>
</tr>
<tr>
<td>7</td>
<td>160</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>590</td>
<td>12</td>
</tr>
<tr>
<td>Mean ± SD</td>
<td>413 ± 160</td>
<td>7.6 ± 3.6</td>
</tr>
</tbody>
</table>

Pdi, transdiaphragmatic pressure.

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Fig. 1. Experimental record of tracings from one patient during spontaneous resting breathing. Raw tracings of EAdi (top), flow (second tracing), esophageal pressure (third tracing), gastric pressure (fourth tracing), and transdiaphragmatic pressure (bottom tracing) in patient with advanced DMD. The EAdi is expressed, as a percentage of the maximum EAdi. Note that cardiac artifacts did not influence the EAdi. Cardiac artifacts or esophageal spasms significantly disturbed the Pes signal.
on the esophageal electrodes during the phrenic nerve stimulation, and the position of the diaphragm could be confirmed along the array by reversal of the CMAPs from one electrode pair to the next [18]. In contrast to what would be expected, our inability to detect the EAdi in patient no. 8 was not related to the weakness (in fact his Pdi sniff was the highest for the group). Hence, our most likely explanation for the lack of detectable signals in this patient is that the diaphragm function around the esophagus may have been more impaired than the more distal areas of the diaphragm.

In the present study, the relative diaphragm activity was more than five times the value observed in healthy subjects [11]. An increase in relative diaphragm activation can either be due to (1) a reduction in maximal pressure generating capacity of the diaphragm due to weakness, (2) an increased load with normal maximal pressure generating capacity (3) a reduced maximal EAdi, or (4) an increased respiratory drive, unrelated to diaphragm weakness or load. DMD patients are likely to be affected by both the severe weakness of the respiratory muscles (reduced maximal pressure generating capacity and reduced maximal EAdi) and increased load.

The loss of diaphragmatic strength in DMD may be due to (i) mechanical weakening of the sarcolemma by cell membrane rupture [19,20], (ii) inappropriate calcium influx (iii) aberrant cell signaling (iv) increased oxidative stress, and (v) recurrent muscle ischemia [21], and/or an altered length–tension relationship due to changes in diaphragm curvature. In general, the disease process can be characterized by a loss of functional motor units. This would reduce both the maximum voluntary EAdi and the maximum pressure generating capacity of the diaphragm. Studies in animal models of DMD show that the diaphragmatic strength is reduced to 25% of control values in older animals [19], down to 13.5% of control at the very late stages of the disease [7]. This was associated with a transition to a slower muscle phenotype (Myosin heavy chain type 1), a moving of the length–tension curve to the left with the optimal length being at 65% of the control value, and a 35% loss in sarcomere number [7]. Recent work in the mdx mouse suggests that contraction-induced injury

Table 3
Dynamic measurements during quiet breathing

<table>
<thead>
<tr>
<th>Patient</th>
<th>VT (ml)</th>
<th>Neural breathing frequency (bpm)</th>
<th>Pdi (cmH2O)</th>
<th>EAdi (%max)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Not available</td>
<td>34</td>
<td>1.7</td>
<td>– (no TLC)</td>
</tr>
<tr>
<td>2</td>
<td>201</td>
<td>30</td>
<td>0.3</td>
<td>20.4</td>
</tr>
<tr>
<td>3</td>
<td>332</td>
<td>22</td>
<td>3.2</td>
<td>35.9</td>
</tr>
<tr>
<td>4</td>
<td>223</td>
<td>26</td>
<td>1.7</td>
<td>62.0</td>
</tr>
<tr>
<td>5</td>
<td>123</td>
<td>32</td>
<td>2.6</td>
<td>61.2</td>
</tr>
<tr>
<td>6</td>
<td>127</td>
<td>18</td>
<td>1.0</td>
<td>62.3</td>
</tr>
<tr>
<td>7</td>
<td>112</td>
<td>24</td>
<td>0</td>
<td>28.6</td>
</tr>
<tr>
<td>8</td>
<td>267</td>
<td>Not available</td>
<td>N/A</td>
<td>–</td>
</tr>
<tr>
<td>Mean±SD</td>
<td>198±83</td>
<td>25±7</td>
<td>1.5±1.2</td>
<td>45.1±19.0</td>
</tr>
</tbody>
</table>

VT, tidal volume; Pdi, transdiaphragmatic pressure; EAdi, diaphragm electrical activity; Max, maximum; Bpm, breaths per minute.

Fig. 2. Experimental record of tracings from one patient during magnetic stimulation. Top panel shows the electrical signals recorded from each electrode pair (ch8 is at the top of the array) and (ch1 is at the bottom of the array) during electrical stimulation. The stimulus artifacts are demonstrated as well as a typical ECG (not to be mistaken as diaphragm electrical activity). The lower panel demonstrates the pressures recorded, and indicate no response of the diaphragm following supra-maximal stimulation.
may be responsible for the muscle weakness [22]. Studies in healthy animals have also indicated that extremely elevated arterial CO2 levels (up to 85 mmHg) may in themselves affect diaphragm contractility [23].

Patients with DMD are expected to have increased loads on the diaphragm due to reduced chest wall compliance, and reduced lung compliance due to microatelectasis [24], scoliosis, and airway secretions caused by ineffective cough from the expiratory muscle weakness. These loads could all potentially demand higher diaphragm activation during resting breathing. Expressed as a percentage of their reserve, however, the $P_{di}$/Pdimax was 18% during quiet breathing.

In the present study, the increased amount of relative diaphragm activation may have been due to the increase in dead space occurring with the addition of the mouthpiece (10 ml) and pneumotach (50 ml). Although it is likely that the patient’s hypercapnia may contribute to the increased neural drive, recent experiments in the mdx mouse indicate a poor/blunted ventilatory response to CO2 [25].

Given the complexity of what factors determine the relative diaphragm activation, it is interesting to observe that the values of relative diaphragm activation found in the present study are comparable to those obtained in two other groups of patients with decreased diaphragm force generating capacity (Fig. 3), where it was demonstrated that patients with post-polio syndrome (neurological disease) and those with COPD (pulmonary, non-neurological disease) also use approximately 40% of their maximum EAad [11]. One similarity between the three patient groups was that PCO2 was around 50 mmHg [11]. With respect to respiratory muscle strength, however, the DMD patients were much weaker as indicated by the $P_{di}$ sniff values (97.6 ± 34.4 cmH2O for the COPD patients, 67.3 ± 31.7 cmH2O for the post-polio patients, and 7.6 ± 3.6 for the patients in the present study). Expressed in relative terms, the maximum volitional pressure generated by the diaphragm in the DMD patients was 12, 8, and 5% of those previously obtained in the COPD, the post-polio patients, and healthy subjects, respectively [11]. Due to this extreme weakness, we would have anticipated larger relative diaphragm activation in the DMD group, however during quiet breathing, the inspiratory $P_{di}$ swings were also lower (1.5 ± 1.2 cmH2O for DMD, 8.1 ± 4.0 cmH2O for COPD, and 7.7 ± 5.3 cmH2O for post-polio).

Considering that the three groups demonstrated similar levels of arterial CO2, although the patients with DMD were considerably weaker than the COPD and Polio patients studied previously [11], we believe that the elevated relative EAad in DMD contradicts the assertion that hypoventilation might be due to a reduced neural drive.

Similar to reports from other authors (e.g. Ref [26]), the patients in the present study demonstrated a rapid shallow breathing pattern and hypercapnia, suggesting that they were adopting this strategy to avoid respiratory muscle fatigue. In fact, the tension-time index ‘TTdi’ was 0.054 (calculated from neural timings) and 0.08 (calculated from flow timings), indicating that their habitual breathing pattern was not in the fatiguing range [27]. Unfortunately, EAad signal quality was not suitable for the power spectrum analysis and reliable measures of CF could not be obtained to infer the early signs of diaphragm fatigue.

One of the possibilities for the hypercapnia is simply that these patients are limited by their diaphragm activation. Recall that diaphragm activation is a term used to describe the combined processes of motor unit recruitment and increased motor unit firing rate. The data from the present study indicates clearly that with 100% recruitment at a firing rate of 1 Hz (i.e. the magnetic stimulation protocol), there was no detectable $P_{di}$ twitch response. This suggests that by recruitment alone, patients with late DMD are not able to generate adequate pressures for ventilation. They must increase the firing rate of the available units. However, it has been demonstrated that an increase in diaphragm activation alone is sufficient to compromise the excitability of the diaphragm fiber membranes, as evaluated by the EAad power spectrum center frequency [28]. Hence, the authors suggest that the mechanism available for these patients to increase their ventilation would be through an increase in diaphragm motor unit firing rate.

It has been reported that patients with advanced DMD have exhibited rhythmic-spontaneous upper-body movements (so called ‘rocking-boat movement’), where the diaphragm descends during inspiration presumably because of the abdomen’s outward movement [29]. In the present study, the occurrence of this rocking-boat motion was not systematically evaluated or quantified. However, we did observe that six out of eight patients had negative gastric pressure swings on inspiration during the quiet breathing period. We can only speculate that this phenomenon would
result in a lowered neural drive to the diaphragm, and hence a lower $\text{EA}_{\text{di}}$ signal.

In the present study, the latency time (time between onset of stimulus and onset of diaphragm CMAP) was $8.2 \pm 1.3$ ms. These results indicate longer latencies than reported in healthy subjects, in whom, at maximal stimulus, the latency was $5.88$ ms with an esophageal electrode (simultaneous surface value $5.68$ ms) [30]. Motor unit remodeling has been observed in younger DMD patients (5–11 years old), where from the electromyogram of single motor unit action potentials, it was suggested that there is a conversion of fast to slow type motor units (innervated by slow non-myelinated fibers) [31].

5. Conclusion

In patients with late DMD who have severely impaired diaphragm function, diaphragm electrical activity was detectable in seven out of eight patients and represented 45% of maximum voluntary activation. The results of this study show that despite the near complete loss of diaphragm force, in advanced DMD, it is still feasible to monitor diaphragm electrical activity with an esophageal electrode array.

References