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Clin Rehabil 2008 22: 741

DOI: 10.1177/0269215508091203

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Comparison study of chest physiotherapy home training programmes on respiratory functions in patients with muscular dystrophy

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Received 20th September 2007; returned for revisions 5th January 2008; revised manuscript accepted 23rd February 2008.

Objective: To compare the effects of home training programmes, threshold inspiratory muscle training and breathing exercise on spirometry and maximal pressures in patients with muscular dystrophy.

Design: Prospective blinded 12-week study.

Settings: Cardiopulmonary department of university hospital.

Subjects: Twenty-three patients with muscular dystrophy (17 patients with limb girdle muscular dystrophy and 6 patients with Becker muscular dystrophy) assigned to the threshold inspiratory muscle training and breathing exercise groups with alternate allocation.

Methods: Spirometry, maximal inspiratory pressure (PI_{max}) and maximal expiratory pressure (PE_{max}) were measured before and after training. In the threshold inspiratory muscle training group threshold pressure load was determined as equal to 30% of weekly PI_{max} measurement. In the breathing exercise group, patients performed deep and forceful diaphragmatic and segmental exercises. All patients performed exercises at home and once a week at hospital under supervision.

Results: The improvement of PI_{max} in the threshold inspiratory muscle training group was more significant than the improvement observed in the breathing exercise group ($P=0.05$). PE_{max} increased significantly only in the breathing exercise group ($P=0.01$). Spirometry results did not change significantly in both groups after the training.

Conclusions: We conclude that respiratory muscle strength is enhanced by training in the patients with muscular dystrophy who are ambulatory, but inspiratory and/or expiratory training effect is specific to the trained muscles. The techniques that improve the strength of respiratory muscles should be included in the physiotherapy management of patients with muscular dystrophy.

Introduction

Muscular dystrophies are progressive, hereditary, degenerative disorders of skeletal muscle. The clinical manifestations of muscle weakness in the different forms of muscular dystrophy

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usually become evident first in the muscles of the limbs and limb girdles.¹ Weakness, which affects proximal limb muscles, may also affect respiratory muscles which may manifest as respiratory distress during the late stages of the disease.² Progressive weakness of the respiratory muscles is one of the major problems in the management of patients with muscular dystrophy and can be at least partially prevented by training of respiratory muscles.^{3,4} The decreasing pulmonary and chest wall compliance observed in neuromuscular diseases increases the mechanical load sustained at each breath by the impaired respiratory muscles. This increased load leads to fatigue and eventual respiratory failure, which is the leading cause of death in these patients.⁵⁻¹⁰ Primary pulmonary insufficiency and failure are associated with severe general disability marking the ultimate stage of the disease.¹¹

Lung volume changes in patients correlate with respiratory muscle weakness in Duchenne muscular dystrophy patients, and although inspiratory muscle dysfunction plays a key role in the development of chronic ventilatory insufficiency, reductions in expiratory muscle strength are the first signs of dysfunction and lead to the first episodes of respiratory failure.¹² In order to prevent the decline in respiratory muscle function, conventional chest physiotherapy techniques such as breathing exercises and various devices have been used.¹³⁻¹⁷ Devices using resistive loads have been developed to train the inspiratory muscles, with the specific goal being to stabilize or improve maximal inspiratory pressure (PI_{max}).¹⁴ In a review it is stated that, training-related improvements in inspiratory muscle performance are more pronounced in patients who are less severely affected by their disease.¹⁵ For this reason, we aimed to evaluate the effects of a respiratory muscle-strengthening programme on respiratory functions in ambulatory patients with limb girdle and Becker muscular dystrophy who have no clinical signs of respiratory problems. The purpose of our study was to compare the effects of two home training programmes – threshold inspiratory muscle training and breathing exercise – on respiratory functions in patients with muscular dystrophy.

Patients and methods

Twenty-three patients with muscular dystrophy (17 patients with limb girdle muscular dystrophy and 6 with Becker muscular dystrophy) were included and assigned to a threshold inspiratory muscle training group (11 patients) or a breathing exercise group (12 patients) with alternate allocation in this study. The diagnosis was made by the neurologist who referred the patients.^{18,19} The inclusion criteria were as follows: (1) being ambulatory; (2) had no visible spinal deformities; (3) had no symptoms or signs of inspiratory muscle fatigue; shortness of breath, orthopnoea or dyspnoea during bathing or swimming, short sentences during speech, tachypnoea, paradoxical movement of abdominal or thoracic wall, problems with cough;²⁰ (4) free from respiratory tract infections, (5) had no symptoms or signs of cardiomyopathy; heart failure symptoms and physical findings. Atrial and ventricular tachyarrhythmias, poor R-wave progression and intraventricular conduction abnormalities, especially left bundle branch block, were not seen in electrocardiography. Besides these, anterior Q waves, ST-segment and T-wave abnormalities and P-wave changes were absent in electrocardiography.

The patients were informed about the study; all agreed to participate and appointments for tests and training education sessions were adjusted to the best time which suited to them.

Study design

In this prospective trial, patients were allocated to either threshold inspiratory muscle training or breathing exercise group alternately according to their arrival order in the hospital (Figure 1). The first patient is allocated to the threshold inspiratory muscle training group and the next one is allocated to the breathing exercise group. With the others, this pattern is preserved. In general, only one patient or no patients arrived at the hospital each week. Once, two patients (sister and brother) arrived on the same day of the week and both were allocated to the breathing exercise group. The range of patients that fulfilled our inclusion criteria for the study per week was 0–2. After the group allocation, patients in both

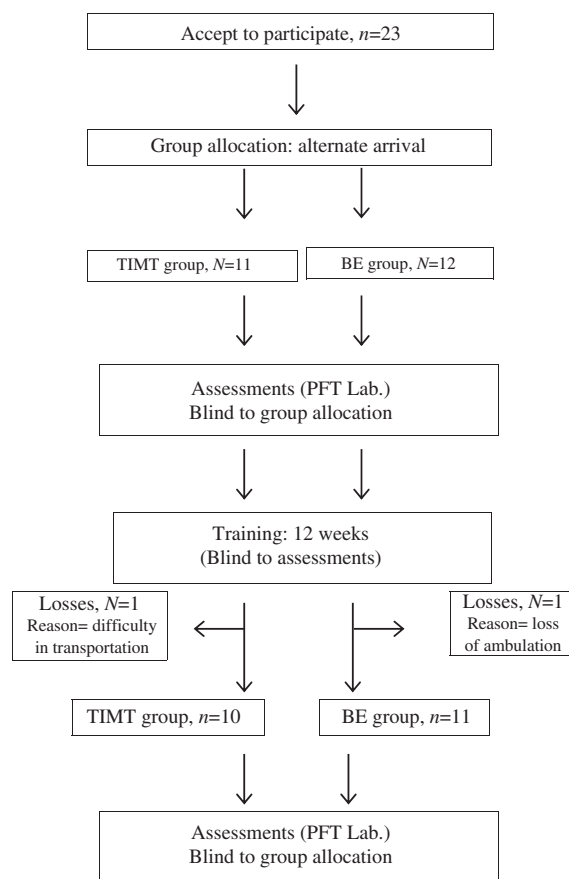


Figure 1 Design of the study. TIMT, threshold inspiratory muscle training; BE, breathing exercise.

groups were evaluated at baseline and after 12 weeks by the same examiner who was blind to group allocation. Both exercise programmes were supervised by the same trainer who was blind to initial and final assessments. We compared the effects of home training programmes (threshold inspiratory muscle training and breathing exercise) on respiratory functions in our patients. The primary analysis of our study was whether there was a greater change in respiratory functions in one group or the other.

Training protocol

In the threshold inspiratory muscle training group, a threshold inspiratory muscle trainer was

used as a training device (Respironics; Cedar Grove, NJ, USA), since the threshold pressure setting is adjustable in $-2\text{ cmH}_2\text{O}$ increments (range, $-7\text{ cmH}_2\text{O}$ to $-41\text{ cmH}_2\text{O}$). The initial training load was 15% of the patient's baseline PI_{max} at first week. In the following weeks, the threshold pressure load was equalized to 30% of weekly PI_{max} measurements as assessed by trainer.

In the breathing exercise group, a physical therapist instructed patients to perform diaphragmatic and segmental exercises.²¹ The patients were encouraged to do deep inspiration and full expiration during all breathing exercises. Exercises were done with 10 repetitions with 30 seconds rest and approximately 8–10 sets were completed in one session.

Patients performed daily threshold inspiratory muscle training or breathing exercise sessions of 15 minutes' duration, twice a day for 5 days per week at home. Our patients were informed about signs of hyperventilation and they were asked to rest after 10 repetitions of exercise to avoid fatigue. In order to control the exercises, to measure mouth pressures and to observe the patients' response to training intensity a supervised training programme was performed in the cardiopulmonary rehabilitation department, once a week in both groups. To promote compliance with the therapy, patients were asked to write a diary of the training periods, which was reviewed weekly. It included the date, time and duration of each training session. The patients were asked to perform threshold inspiratory muscle training or breathing exercise at the same time each day, so that a daily routine habit could be established.

Outcome measures

Spirometric measures

Spirometric measures were performed using a dry spirometer (Vitalograph, Buckingham, UK) and the measured parameters were vital capacity (VC), forced vital capacity (FVC), forced expiratory volume in one second (FEV_1).²² Pulmonary function values were based on the best of three efforts. Measured parameters were converted into predicted value percentages using Kamburoff–Voitovits nomogram.²³

Respiratory strength

Maximal inspiratory and expiratory muscle strength was measured with Sensor MEDICS Micro MPM device (normal range 70–100 cmH₂O and 90–140 cmH₂O, respectively). Measurements were performed using the technique of Black and Hyatt.²⁴ PI_{\max} was measured after full expiration and maximal expiratory pressure (PE_{\max}) after full inspiration.²⁵ All subjects had three to five trials in order to negate possible learning effect. Assessments were done until three technically satisfactory and consistent values were obtained and the highest value was used in data analysis. Each subject wore a nose clip and breathed through a mouthpiece.

All measurements were performed in seated position at baseline and after the 12-week training by the same examiner who was blind to group allocation.

Statistical methods

Data are presented as mean and SD. Mann–Whitney *U*-test was used to compare the differences between the two groups. Within groups, baseline and post training results were compared using Wilcoxon test. The level of significance was set at 0.05.

Results

Patient characteristics of two groups were similar and there was no significant difference between the baseline values of mouth pressure and spirometry (Table 1). Two subjects (one from each group) dropped the study because of difficulty in transportation and loss of ambulation. Twenty-one patients completed the study. The mean (SD) age was 22.50 (7.50) years in the threshold inspiratory muscle training group, and 24.27 (9.40) years in the breathing exercise group. Two patients smoked in the threshold inspiratory muscle training group, and three smoked in the breathing exercise group.

The comparison of pre–post training difference in mouth pressure and spirometry measurements is summarized in Table 2. The pre–post training difference of PI_{\max} in the threshold inspiratory

Table 1 Comparison of patient characteristics between two groups

Parameters	TIMT group	BE group	<i>P</i> -value
Male:female	7(70%):3(30%)	6(55%):5(45%)	NS
Age (years)	22.50 ± 7.50	24.27 ± 9.40	NS
Disease duration (years)	11.50 ± 4.97	14.55 ± 9.59	NS
Height (cm)	162.50 ± 13.50	159.18 ± 10.16	NS
Weight (kg)	57.80 ± 14.49	50.5 ± 11.11	NS
PI_{\max} (cmH ₂ O)	85 ± 35.80	68.54 ± 32.35	NS
% PI_{\max}	93.90 ± 34.32	80.18 ± 32.27	NS
PE_{\max} (cmH ₂ O)	65.70 ± 25.58	65.18 ± 22.99	NS
% PE_{\max}	54.10 ± 19.89	56.18 ± 17.89	NS
VC (L)	3.46 ± 1.12	3.24 ± 0.86	NS
FVC (L)	3.50 ± 1.16	3.30 ± 0.87	NS
% FVC	84.30 ± 16.24	86.72 ± 20.49	NS
FEV ₁ (L)	3.30 ± 1.13	3.13 ± 0.76	NS
% FEV ₁	91 ± 18.79	88.90 ± 31.41	NS

Data are presented as *n* (%) or mean ± SD.

TIMT, threshold inspiratory muscle training; BE, breathing exercise; SD, standard deviation; PI_{\max} , maximal inspiratory pressure; PE_{\max} , maximal expiratory pressure; VC, vital capacity; FVC, forced vital capacity; FEV₁, forced expiratory volume in one second; NS, not significant.

muscle training group was significantly greater than that in the breathing exercise group ($P=0.05$). There was significant improvement in PE_{\max} only in breathing exercise group after the training ($P=0.01$). There was no statistically significant difference between the baseline and after 12-week training results of spirometry in both groups.

Discussion

In our study, significant increases in respiratory muscle strength were achieved after 12 weeks of home training in both threshold inspiratory muscle training and breathing exercise group in patients with limb girdle muscular dystrophy and Becker muscular dystrophy. The pre–post training difference of PI_{\max} in the threshold inspiratory muscle training group was significantly greater than the pre–post training improvement observed in the breathing exercise group. Besides, we found significant improvement in PE_{\max} only in the breathing exercise group after the training.

Table 2 Comparison of changes in mouth pressure measurements and spirometry results before and after training

Variables	TIMT group		BE group		Between groups <i>P</i> -value
	Mean \pm SD	<i>P</i> -value	Mean \pm SD	<i>P</i> -value	
AT PI_{\max} -BT PI_{\max} (cmH ₂ O)	37.50 \pm 22.75	<i>P</i> <0.01	19.00 \pm 16.70	<i>P</i> <0.01	<i>P</i> =0.05
AT PE_{\max} -BT PE_{\max} (cmH ₂ O)	7.80 \pm 14.18	<i>P</i> >0.05	10.27 \pm 12.14	<i>P</i> =0.01	<i>P</i> >0.05
AT VC-BT VC (L)	0.04 \pm 0.14	<i>P</i> >0.05	0.06 \pm 0.17	<i>P</i> >0.05	<i>P</i> >0.05
AT FVC-BT FVC (L)	0 \pm 0.10	<i>P</i> >0.05	-0.01 \pm 0.17	<i>P</i> >0.05	<i>P</i> >0.05
AT FEV ₁ -BT FEV ₁ (L)	0.04 \pm 0.11	<i>P</i> >0.05	0.01 \pm 0.17	<i>P</i> >0.05	<i>P</i> >0.05

TIMT, threshold inspiratory muscle training; BE, breathing exercise; SD, standard deviation; AT, after training; BT, before training; PI_{\max} , maximal inspiratory pressure; PE_{\max} , maximal expiratory pressure; VC, vital capacity; FVC, forced vital capacity; FEV₁, forced expiratory volume in one second.

The results of our study showed that respiratory muscle strength is enhanced in ambulatory patients with muscular dystrophy, but training effect is specific to the trained muscle.

In our study, results of spirometry did not change significantly after training in both groups. This finding was not unexpected since pretraining assessment results of spirometry of our patients were normal. These findings were in agreement with their clinical condition since they were ambulatory and have no clinical signs of respiratory problems. Only their maximal expiratory mouth pressures were below normal values, and this finding clearly shows that reduction in muscle strength occurs while their respiratory function is still maintained. Another point is that although they are ambulatory, this reduction in expiratory muscle strength may be the first sign of deterioration of respiratory function, as it is in patients with Duchenne muscular dystrophy. So it is important to evaluate maximal mouth pressures in all patients with limb girdle and Becker muscular dystrophy routinely without waiting for clinical deterioration. Gigliotti *et al.*²⁶ also reported that the respiratory muscles, especially expiratory ones, are weak in patients with limb girdle dystrophy. The maximum expiratory pressure was the most sensitive indicator of weakness and was decreased in 87% of adult patients with myotonic dystrophy, myasthenia gravis, amyotrophic lateral sclerosis and Duchenne dystrophy.²⁷ According to the initial assessments, our study showed that expiratory muscle strength decreased by 46% in the threshold inspiratory muscle training and 44% in the breathing exercise group.

Despite methodological differences among studies, investigators have generally shown that inspiratory muscles are similar to other skeletal muscle groups in that they can be trained for both strength and endurance in these patients.²⁸⁻³⁰ Koessler *et al.*¹⁶ showed that with inspiratory muscle training, respiratory muscle function could be improved in the long term of up to two years. In the literature, resistive training has been used in general to improve respiratory muscle strength or endurance. Dimarco *et al.*¹⁷ concluded that inspiratory resistive training improves respiratory muscle endurance in muscular dystrophy patients. They stated that improvement in respiratory muscle function may serve to delay the onset of respiratory complications in patients with muscular dystrophy. The authors also found that the degree of improvement with training was directly related to the patients' baseline vital capacity. In agreement with the findings of Dimarco *et al.*, Wanke *et al.*¹⁴ reported that the patients with the most severely reduced vital capacities responded less or did not respond to training in their randomized study. In addition they concluded that specific inspiratory muscle training is useful in the early stages of Duchenne's muscular dystrophy when the vital capacity is better preserved. In our study, vital capacity values of our ambulatory patients were within the normal reference range and only a few patients had borderline pathologic values, besides they had no inspiratory muscle weakness (baseline PI_{\max} 93.90 cmH₂O in the threshold inspiratory muscle training group and 80.18 cmH₂O in the breathing exercise group). At the end of the 12-week training we

had significant improvement in our patients' respiratory muscle strength whose respiratory functions were well preserved. This supports the findings of Wanke *et al.*¹⁴ and Dimarco *et al.*¹⁷

Topin *et al.*²⁹ suggested that specific training improves respiratory muscle endurance in Duchenne muscular dystrophy and the effectiveness of training appears to be dependent on the quantity of training. Winkler *et al.*¹³ reported that in patients with a decline of less than 10% of vital capacity in the year before training, the desired effects of inspiratory muscle training are dose-dependent. In our study, the threshold inspiratory muscle training group, in which training intensity increased weekly, showed higher improvements than the breathing exercise group. For this reason, we agree with Topin *et al.*²⁹ and Winkler *et al.*¹³ that the effects of threshold inspiratory muscle training are dose-dependent.

Most authors focused on resistive training but easily applicable breathing exercises were not investigated in muscle training studies. In the literature there is only one study investigating the effects of breathing exercises in patients with muscular dystrophy. Ugalde *et al.*³⁰ found that pursed lips breathing and deep breathing are effective and easily employed strategies that significantly improve tidal volume and oxygen saturation in subjects with myotonic muscular dystrophy. But the effects of breathing exercise on respiratory functions were not evaluated. In our study, we observed improvement in inspiratory muscle strength with both daily resistive inspiratory muscle training and unresistive breathing exercises. We think that the underlying mechanism of improved inspiratory and expiratory muscle strength of patients in the breathing exercise group is the result of the effect of deep and forceful inspiration and expiration exercises on muscles.

In our study, although there were no significant differences in spirometric measurements, the increase in muscle strength in both groups may have positive effects on lung function and may stabilize or at least slow down the decline in vital capacity by threshold inspiratory muscle training or breathing exercises. For this reason, the techniques that improve the strength of respiratory muscles should be included in the physiotherapy management of patients with muscular dystrophy.

In addition to objective findings, the compliance to the training of our patients in both groups was excellent. There was no patient drop-out from the study because of unwillingness to continue the training. Our patients had difficulty in transportation and reaching to hospital independently was quite hard. So our home training programme was suited well to our study population. We also think that monitoring the patients' diaries promoted compliance with the therapy.

One weakness of our study was the lack of randomization; we divided our patients into groups with alternate allocation. Another limitation is that some patients did not agree to participate in this study because they had transportation problems and for this reason our study groups were small.

We did not include expiratory muscle strengthening exercises in addition to inspiratory training in the threshold inspiratory muscle training group. The results of our study (improvement in PI_{max} in the threshold inspiratory muscle training group and both PI_{max} and PE_{max} in the breathing exercise group) suggested that if we did so, we may have had improvement in expiratory muscle strength also. Further studies are needed to investigate two-way training effect in these patient groups.

From the present study we concluded that whether resistive or not the outcome may be better if the training programme includes both inspiratory and expiratory training. We recommend inclusion of approaches that improve respiratory functions in the general rehabilitation programmes in ambulatory patients with muscular dystrophy, even if they have no clinical signs of respiratory problems.

Clinical messages

- Inspiratory muscle strength can be increased in patients with muscular dystrophy with and without resistive breathing exercises.
- Expiratory muscle strength can be increased by deep and forceful breathing exercises.
- There were no side-effects during the training, techniques were simple, and could be carried out easily at home.

Acknowledgements

This study is partially supported by Istanbul University Research Foundation (Project T-888/17072000).

The authors gratefully thank Professor Coskun Ozdemir, from the Neuromuscular Disorder Association of Turkey and Professor Piraye Serdaroglu from Istanbul University, Istanbul Medical Faculty, Department of Neurology for patient recruitment, and the physical therapists of the Pulmonary Function Test Laboratory at Istanbul University, Cardiology Institute for spirometry and respiratory muscle strength assessments of patients.

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