2 Years' Experience With Inspiratory Muscle Training in Patients With Neuromuscular Disorders*

Wolfgang Koessler, MD; Theodor Wanke, MD; Guenther Winkler, MD;
Astrid Nader, MD; Karl Toifl, MD; Herbert Kurz, MD; and
Hartmut Zwick, MD, FCCP

Purpose: The aim of our study was to assess the long-term effects of specific inspiratory muscle training (IMT) in patients with neuromuscular disorders (NMDs) who have various degrees of respiratory impairment.

Patients and methods: Twenty-seven patients with NMDs (Duchenne's muscular dystrophy, 18 patients; spinal muscular atrophy, 9 patients) underwent 24 months of IMT. Patients were divided into three groups according to their vital capacity (VC) values. VC was measured as the parameter for the respiratory system involvement of the disease. Maximal inspiratory pressure (PImax) was assessed as the parameter for respiratory muscle strength, and the results of the 12-s maximum voluntary ventilation test (12sMVV) were assessed as the parameter for respiratory muscle endurance. Pulmonary and inspiratory muscle function parameters were assessed 6 months before training, at the beginning of training, and then every 3 months.

Results: The PImax values improved in group A (VC, 27 to 50% predicted) from 51.45 to 87.00 cm H_2O, in group B (VC, 51 to 70% predicted) from 59.38 to 94.4 cm H_2O, and in group C (VC, 71 to 96% predicted) from 71.25 to 99.00 cm H_2O. The 12sMVV values improved in group A from 52.69 to 69.50 L/min, in group B from 53.18 to 62.40 L/min, and in group C from 59.48 to 70.5 L/min. For all three groups, there was a significant improvement of PImax (p < 0.007) and 12sMVV (p < 0.015) until the 10th month when a plateau phase was reached with no decline in the following month until the end of training.

Conclusion: With IMT, respiratory muscle function can be improved in the long term of up to 2 years. *(CHEST 2001; 120:765–769)*

Key words: inspiratory muscle training; long-term effects; neuromuscular disorders

Abbreviations: IMT = inspiratory muscle training; 12sMVV = 12-s maximum voluntary ventilation; NMD = neuromuscular disorder; PImax = maximal inspiratory mouth pressure; VC = inspiratory vital capacity

Inspiratory muscle function impairment causes an increased risk of respiratory complications and premature death in patients with neuromuscular disorders (NMDs).[1] However, it has been shown that in healthy subjects and in patients with pulmonary diseases, kyphoscoliosis, or Duchenne's muscular dystrophy, respiratory muscles and, in particular, the diaphragm are trainable in terms of strength and endurance.[2–7] But despite these results, inspiratory muscle training (IMT) is controversial in rehabilitation programs for NMD patients. It may be that no studies have assessed the effects of IMT over a period of > 6 months.

The goal of our study was to answer the following questions:

1. What are the effects of IMT after long-term training of 24 months on the inspiratory muscle function and lung function parameters, and is it possible to counteract the natural decline of vital capacity (VC) in NMD patients?
2. Is the long-term effect of IMT affected by the severity of the respiratory system involvement in NMDs?
3. Is the motivation to train (*ie*, the number of correctly performed exercises) influenced by the severity of the disease?

Group	Age, yr	VC L	VC % Predicted	FEV_1 L	FEV_1 % Predicted
A	19.91 ± 4.99	1.46 ± 0.54	42.6 ± 19.7	1.33 ± 0.49	53.1 ± 21.0
B	16.45 ± 4.03	1.91 ± 0.56	62.4 ± 20.1	1.60 ± 0.53	64.3 ± 22.8
C	12.55 ± 2.28	2.22 ± 0.70	78.7 ± 18.2	1.96 ± 0.59	75.5 ± 24.9

*Values are given as mean ± SD.

Materials and Methods

Subjects

Twenty-seven patients with NMDs (Duchenne's muscular dystrophy, 18 patients; spinal muscular atrophy, 9 patients) participated in our study. None of these patients had participated in our previous study, which was published in 1994.[2] Before entering the study, patients were divided into the following three groups according to their VC values, which served as the parameter for the respiratory system involvement of the disease: group A patients had a VC range of 27 to 50% predicted; group B patients had a VC range of 51 to 70% predicted; and group C patients had a VC range of 71 to 96% predicted. Each group consisted of nine patients, including three patients with spinal muscular atrophy. The baseline data are shown in Table 1. The mean (± SD) age of all patients (N = 27) was 16.30 ± 4.84 years. All patients over 10 years of age were wheelchair bound.

We have previously demonstrated[2] that in patients with VC values < 25% predicted, specific training of the inspiratory muscles does not produce any benefit. So, in this study the baseline VC values at the beginning of IMT were at least 27% predicted. We used VC as a parameter for the progression of disease involving the entire respiratory system, because it is affected not only by respiratory muscle capacity, but also by chest wall and lung compliance.[8]

The diagnosis of the disease was made between 3 and 5 years of age on clinical, enzymatic, electromyographic, and muscle biopsy criteria. All patients were free from respiratory tract infections and had no symptoms or signs of inspiratory muscle fatigue, which are the obligatory indications for rest.[9] No patient complained of sleep disturbance, daytime hypersomnolence, or morning headaches, and none had had an episode of acute respiratory failure requiring endotracheal ventilation. Capillary blood collected from an earlobe was used to analyze blood gases. No patient was hypercapnic, and all had normal Po_2 values. Informed consent was obtained from all subjects, and the study was approved by the Human Subject Committee of the City Hospital Lainz.

Training Apparatus

The training apparatus (E. Biegler Co; Mauerbach, Austria) that we used and the training itself were described in detail in our previous study.[2] Each patient had his own training apparatus and trained at home. It enabled the patient to do resistive breathing maneuvers against variable inspiratory resistance for endurance training or maximal static inspiratory efforts against almost occluded resistance for strength training. Pressure was measured with a ± 300 cm H_2O differential pressure transducer (model 142 PC; Honeywell; Freeport, IL). To provide a visual control of the performance, we used a visual feedback system. During endurance training, there was a light-emitting diode display whereby the patients were shown breath by breath the inspiratory airflow actually achieved in arbitrarily chosen units, and they were shown the minimal inspiratory flow they had to reach during each inspiration. Patients were also shown whether they had achieved the minute ventilation value level while breathing against the inspiratory resistance. During the 1-min breathing cycle, if the minimal airflow value was not achieved more than twice, a warning signal on the training apparatus would be shown to the patient, and the cycle had to be repeated. The same happened if the desired minute ventilation level was not achieved.

During muscle strength training, the visual feedback system shows the minimal inspiratory pressure that had to be achieved and the actual inspiratory pressure that was reached.

Lung Function and Respiratory Muscle Function Parameters

Ventilatory function and inspiratory muscle function were evaluated 6 months before training, at the beginning of the training, in the first month of training, then every 3 months, and at the end of training after 24 months. We used the 12-s maximum voluntary ventilation (12sMVV) test as the parameter for respiratory muscle endurance, and maximal inspiratory mouth pressure (PImax) as the parameter for respiratory muscle strength. All measurements were made with the patient in the sitting position. The VC and the 12sMVV test were measured three times on a computerized spirometer (Jaeger; Würzburg, Germany). The best trial for the 12sMVV test was used for further analysis. The patient used a flanged mouthpiece, which was held in the mouth behind the lips and firmly gripped by the teeth. This mouthpiece also was used for training at home.[10] The best of 10 PImax maneuvers was used for the analyses.

The baseline lung function parameters of the three patient groups are shown in Table 1. The expected normal values are those reported by Quanjer et al.[11] In patients with scoliosis, arm

Group	6 mo Before Training, cm H_2O	Beginning of Training, cm H_2O	Plateau Phase, cm H_2O	End of Training, cm H_2O
A	54.41 ± 24.56	51.45 ± 20.67	79.43 ± 27.73	87.00 ± 12.73
B	61.80 ± 18.5	59.38 ± 19.45	92.00 ± 12.9	94.4 ± 29.94
C	72.28 ± 23.34	71.25 ± 22.87	97.88 ± 25.94	99.00 ± 26.87

*Values are given as mean ± SD.

Table 3—*12sMVV Values During the Observation Period**

Group	6 mo Before Training, L/min	Beginning of Training, L/min	Plateau Phase, L/min	End of Training, L/min
A	53.23 ± 27.8	52.69 ± 30.48	65.66 ± 31.71	69.50 ± 21.75
B	56.9 ± 24.75	53.18 ± 22.94	60.04 ± 27.08	62.40 ± 28.35
C	61.2 ± 14.2	59.48 ± 16.13	65.13 ± 18.4	70.50 ± 10.61

*Values are given as mean ± SD.

span was used to determine the percent predicted values according to the method of Johnson and Westgate.[12] PImax was measured by using the training apparatus. Ten maximal static inspiratory efforts against the almost occluded resistance were performed. An interval of 30 to 40 s was allowed between each maneuver, and the highest pressure obtained was used for further analyses.

Training Protocol

At home, patients had to perform both resistive breathing maneuvers and maximal static inspiratory efforts. Training was performed with the patient in the sitting position, and a noseclip was used. Inspiratory resistive breathing training consisted of 10 loaded breathing cycles of 1-min duration each, with 20-s intervals between them. The level of the inspiratory resistance was adjusted in the hospital every 3 months so that 70 to 80% of the PImax value had to be generated with each breath. Thus, each patient had to correctly complete 10 resistive breathing cycles twice a day. The number of correctly and incorrectly performed exercises was stored by the apparatus to enable the physician to check the training of the patient. An incorrect test result represents a failure of the patient to achieve the target pressure or flow levels, or a failure to perform the total number of exercise cycles at all. Fifteen minutes before the resistive breathing training, the patients had to perform 10 maximal static inspiratory efforts and reach a certain minimal pressure value, which was at least 90% of the maximally generated inspiratory pressure that had been determined in the hospital. This value also was determined and was adjusted in the hospital in each follow-up. A 20-s interval between the maneuvers was allowed. If the minimal pressure values were not achieved, this maneuver had to be repeated until a total of 10 maneuvers were performed correctly.

Patients had to perform strength and endurance training twice daily, but if they could not perform the prescribed number of exercises, they were not excluded from our study.

Statistical Analysis

We used the rank-sum test for statistical analyses, a nonparametric test to check the median differences between different groups or different points of time.

Results

Baseline values are shown in Table 1. Table 2 shows the PImax values 6 months before training, at the beginning of training, at the plateau phase of training, and at the end of training. For all groups, there was a significant improvement of PImax ($p < 0.007$) until the 10th month when a plateau

phase was reached. Table 3 shows the 12sMVV values 6 months before training, at the beginning of training, at the plateau phase of training, and at the end of training. There was also a significant increase in 12sMVV ($p < 0.015$) until the 10th month when a plateau phase was reached. This means that there was a significant improvement in muscle strength and endurance time during the first 10 months and that after that time the improvement in PImax and 12sMVV was much slighter.

In all three groups, we could find no significant decrease in VC values during training (Table 4) and no significant difference in the number of correctly performed maneuvers. The mean number of correctly performed exercises in strength and endurance training within 3 months is shown in Table 5.

Discussion

Our study yielded the following results:

1. Respiratory muscle strength and endurance can be improved by long-term training in patients with VC values > 25% of those predicted.
2. There was no significant decline in VC after a training period of 24 months.
3. It is possible to motivate young patients to train at home, irrespective of their lung function impairment.

As far as we know, this is the first study that analyzed the long-term effects of a 24-month IMT program in patients with NMDs. During this time, periodic measurements of lung function parameters and respiratory muscle function parameters, like inspiratory muscle strength and endurance, were performed.

Table 4—*VC Values During the Observation Period**

Group	6 mo Before Training, L	Beginning of Training, L	End of Training, L
A	1.57 ± 0.57	1.46 ± 0.54	1.74 ± 0.60
B	1.98 ± 0.52	1.91 ± 0.56	1.94 ± 0.58
C	2.24 ± 0.76	2.22 ± 0.70	2.35 ± 0.84

*Values are given as mean ± SD.

Table 5—*Mean Number of Correctly and Incorrectly Performed Exercises Within 3 mo*

	Strength Exercises		Endurance Exercises	
Group	Correct Trials	Incorrect Trials	Correct Trials	Incorrect Trials
A	116.8 ± 19.7	3.9 ± 2.2	106.3 ± 16.7	7.7 ± 2.9
B	113.1 ± 15.9	4.3 ± 2.3	111.1 ± 11.0	6.6 ± 3.6
C	112.7 ± 20.3	3.2 ± 1.4	109.7 ± 25.4	5.0 ± 2.6

*Values are given as mean ± SD.

One of the main problems of NMDs is the progressive impairment of inspiratory muscle function. This leads to fatigue and eventually to respiratory failure, which is the most important cause of death in these patients.[13] Therefore, it seems logical to use respiratory muscle training to improve inspiratory muscle function and to delay the early onset of respiratory failure in patients with NMDs. In the literature, the role of IMT has been controversial in patients with NMDs. In some studies[2,6,14] in which inspiratory resistive loaded breathing was used, muscle strength and endurance increased. But in these studies, the training period often lasted not more than a few weeks, so there is no evidence that these effects would continue over a long period. In other studies,[15,16] only endurance increased, and there was no improvement in muscle strength. The reason could be that the intensity of training was too low, because there was no control of training intensity. Respiratory muscles seem to exhibit similar adaptations to training to those of other skeletal muscles, provided that the training follows the basic training principles for any other striated muscle with regard to intensity, duration of the stimulus, and the specifics of the training.[17] However, it also has been argued[18] that IMT is hazardous since it may accelerate fatigue in the respiratory muscles by overwork.

We could show that IMT, combined with visual feedback, improved respiratory muscle strength and endurance even in the long term, provided that the patient's ventilatory function was not severely impaired.[2] The visual feedback allows proper monitoring of the training intensity, so that sufficient training could be carried out and its positive effects on respiratory muscles could be explained.

Although the advantage of improved inspiratory muscle function is obvious, it is certainly not easy to motivate especially young patients for the training. Respiratory muscle training is considered cumbersome, boring, and without immediate reward, so that it is very unlikely that many patients will use it long enough to reap the benefits.[19] As a reward, our training equipment identifies incorrect exercises, so that they can be repeated without much waste of time and patients can complete their exercises quickly. A further advantage is that training can be done at home without abandoning close monitoring of the exercises. This complies with the ideal that patients with NMDs should be treated as outpatients, while keeping admissions to the hospital to a minimum.[20] In our three patient groups, there was no difference in the number of correctly performed maneuvers in strength and endurance training, which means that the motivation to perform IMT is not influenced by the severity of the disease.

In addition to improvement in respiratory muscle function, there were also positive effects on lung function. In previous studies, it has been shown that during the growth phase VC increases normally and is identical to its predicted value.[21] The plateau phase generally occurs between 10 and 12 years of age when the VC stabilizes, and then VC decreases as the disease progresses and is complicated by orthopedic deformities.[21]

It has been a great challenge to stabilize VC, or at least to retard its decline by IMT. We have shown that IMT leads to a stabilization of VC, which is important because VC is regarded as a predictor of the need for mechanical ventilation.[22] It may be argued that PImax and 12sMVV test results increase due to a learning effect rather than indicating a real increase in strength and endurance.[14,17] However, an increase due to learning should be most marked between the initial measurements, but such a change was not found. There was a continuous rise in 12sMVV test results in all patient groups until the plateau was reached after 10 months.

The results of our study have shown beneficial effects of respiratory muscle training on 12sMVV test results and PImax measurements during a 2-year period. Therefore, long-term IMT in patients with NMDs is justified.

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