

# Improved Pulmonary Function and Exercise Tolerance With Inspiratory Muscle Conditioning in Children With Cystic Fibrosis\*

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This study documented the effect of inspiratory muscle conditioning in children with cystic fibrosis. Subjects, ages 7 to 14 years, were divided into two groups. The experimental group (n = 10) trained at a high pressure load ( $\geq 29$  cm H<sub>2</sub>O) and the control group (n = 10) trained at a minimal pressure load ( $\leq 15$  cm H<sub>2</sub>O), using a threshold loading device. Subjects trained 30 min a day for 10 weeks. Pulmonary function, inspiratory muscle strength, and exercise tolerance were measured at the beginning and end of the training period. Pulmonary function was measured by body plethysmography. Inspiratory muscle strength was determined by standard measures of maximal inspiratory

Respiratory dysfunction in patients with cystic fibrosis (CF) is associated with hyperinflation, increases in airway resistance, and elevated work of breathing.<sup>1-3</sup> These changes may compromise respiratory muscle function, making the patient with CF more susceptible to ventilatory failure<sup>4</sup> or ventilatory limitation during exercise,<sup>5</sup> particularly late in the course of the disease. However, much of what is currently known about the effects of hyperinflation and increased work of breathing on the respiratory muscles has come from studies on adults with chronic obstructive pulmonary disease (COPD);<sup>3</sup> less is known about patients with CF.

Despite the similarities between the two conditions, there is some controversy as to the extent to which the respiratory muscles are dysfunctional in patients with CF. For example, some investigators have reported relatively well-preserved inspiratory muscle strength,<sup>6</sup> particularly when expressed in relation to lung volume<sup>7-9</sup> while others have shown that it is diminished.<sup>4,10,11</sup> A reduction in maximum inspiratory muscle strength may be most evident in severely hyperinflated or malnourished subjects.<sup>12</sup> In general, respiratory muscle endurance is believed to be elevated in patients with CF,<sup>13,14</sup> but this is also complicated to some extent by the fact that endurance is estimated relative to strength or ventilatory capacity, which can appear increased or diminished depending on the patient's functional status.

Considering the fact that many patients with CF

pressure against an occluded airway. Exercise tolerance was measured by the length of time subjects could walk on a treadmill. Findings indicated that the experimental group showed significant increases in inspiratory muscle strength, vital capacity, total lung capacity, and exercise tolerance in comparison to the control group. (*Chest* 1993; 104:1490-97)

CF = cystic fibrosis; IC = inspiratory capacity; IMT = inspiratory muscle training; MVV = maximal voluntary ventilation; NIH = National Institutes of Health; P<sub>imax</sub> = maximal inspiratory mouth pressure

appear to have a relatively high functional status of their respiratory muscles compared with normal individuals, is there any reason to believe that they would benefit from respiratory muscle training? Gaultier et al<sup>3</sup> (1985) have suggested that the work of breathing is greatly elevated at rest in some patients with CF, approaching a level that would induce fatigue and/or failure of the respiratory pump. Furthermore, Asher et al<sup>6</sup> demonstrated indirect evidence of fatigue in 8 of 11 patients at two thirds of maximum work capacity. Changes at this level could be consistent with impending respiratory muscle fatigue. Therefore, it is possible that the apparently high functional status of the respiratory muscles in some patients with CF may be needed to overcome the elevated impedance of their respiratory system and that additional respiratory muscle training may be beneficial. Two studies have attempted respiratory muscle training in patients with CF using sustained isocapnic hyperpnea<sup>14</sup> or resistive breathing.<sup>7</sup> Although both studies demonstrated improvements in respiratory muscle performance in their patients, the extent to which this resulted in an improvement in the patients' overall functional status was not clear.

In this study, we chose to reexamine the potential influence of inspiratory muscle training (IMT) in young children with CF. The rationale for addressing this problem is that more reliable methods have recently become available which allow for a larger and better controlled conditioning stimulus. The method has recently proved effective in the adult COPD population,<sup>15,16</sup> but has not been applied to patients with CF. A second aim of this investigation was to evaluate whether an IMT program could be effectively incorporated into the physical therapy program of a group

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of relatively young patients. To our knowledge, there have been no studies that have addressed these aims in a young population. Because of the rapid progression of the disease over the second and third decades of life, improvement in inspiratory muscle capacity to normal or supernormal levels early in the time course of the disease may be potentially useful in attenuating the decline in functional status later in time.

The results of this study suggest that an effective training program can be established in young children with CF. Using comparisons between a group undergoing high-intensity training vs a control group undergoing low-intensity "sham" training, significant improvements in inspiratory muscle strength, lung volumes, and exercise capacity are demonstrated over a 10-week training period.

## METHODS

### Subjects

Children with CF ( $n = 20$ ) ranging in age from 7 to 14 years were recruited from a large midwestern CF clinic. Severity of disease was evaluated with a scoring system devised at the National Institutes of Health (NIH) by Taussig et al.<sup>17</sup> These measures, called NIH scores, ranged from 96 to 67 percent in this population (mean = 87.1 percent). The NIH scoring system provides an estimate of the severity of illness of patients with CF with 100 percent being the best possible score. Points are deducted based on evaluation of pulmonary radiograph, pulmonary function tests and related pulmonary complications, patient weight, activity, and attitude. The patients were also evaluated for sexual maturity using the Tanner score, with 4 representing the highest degree of sexual maturity and 1, the least.<sup>18</sup>

The protocol was approved by the Human Subjects Committee of The Ohio State University and Columbus Children's Hospital and consent was obtained from all subjects and families. The children were randomly assigned to an experimental or a "sham" control group.

Prior to data collection, the subjects went through two training and instruction periods in which they were taught how to breathe with the threshold loading device (*ie*, Threshold, HealthScan, Inc) and how to perform maximal inspiratory mouth pressure (P<sub>imax</sub>) maneuvers. The threshold loading device is a hand-held tube with a spring-loaded valve that provides a constant and specific pressure load, independent of inspiratory air flow. In two of the older children in the experimental group, a larger spring with approximately 2× the tension was substituted for the manufactured spring in order to increase the load.

### Training Procedures

Experimental subjects trained at pressure loads up to 60 percent P<sub>imax</sub>, whereas the control subjects trained at 10 percent P<sub>imax</sub>, a light load that will not induce improvement.<sup>15</sup> Subjects were instructed to use the trainer for 30 min a day, 7 days a week, for 10 weeks. Home visits were made to each child for supervision of the training sessions, three times a week, by a nurse. The child was monitored using the training device for 30 min and encouraged to inspire deeply, to near total lung capacity (TLC) with each breath. The children were instructed to use the trainer the other 4 days of the week, without supervision of the investigative team. However, most were supervised by parents on these days. To ensure compliance, the children were encouraged to place stickers on a diary each time they used the trainer for the full 30 min.

Initially, the threshold loading device was set to a low tension of

approximately -7 cm H<sub>2</sub>O. In the experimental group, the spring tension was increased weekly as long as the child could tolerate taking full breaths for the entire 30 min. In general, the pressure was increased to approximately 50 to 60 percent of initial P<sub>imax</sub>. The tension on the control group was increased slowly and minimally so that the maximal tension of the spring was no more than 10 percent of their P<sub>imax</sub> at any time. In all other ways, the controls were handled identically to the experimental subjects.

### Prestudy and Poststudy Evaluation

Inspiratory muscle strength was evaluated from maximum inspiratory pressure measurements against an occluded airway at functional residual capacity (FRC) using standard techniques.<sup>4,19,20</sup> A portable pressure gauge was used to measure P<sub>imax</sub> (Magnahelic model 2000-200-CM, Dwyer Instruments Inc, Michigan City, Ind). The highest pressure achieved after three attempts was recorded. Prior to prestudy data collection, the patients practiced on two occasions to perform the maneuvers (three P<sub>imax</sub> tests per session). Therefore, each subject had six or more practice P<sub>imax</sub> maneuvers prior to the prestudy P<sub>imax</sub>.

An attempt was made to evaluate the maximum work capacity of the inspiratory muscles using an incremental loading procedure. The inspiratory pressure load was produced by a device similar to that of Nickerson and Keens<sup>15</sup> and could operate at higher pressures than the valves used for training. Weights on the threshold valve were changed after every four breaths in approximately 25-g increments until the child could no longer lift the weight. Breathing pattern was controlled during the endurance testing by means of a tape-recorded message simulating the correct breathing patterns. Unfortunately, many of the children did not consistently follow directions on this testing procedure and therefore the results were not believed to reflect their inspiratory muscle work capacity and are not reported herein.

Maximal exercise testing was accomplished on a treadmill using the Bruce protocol.<sup>21</sup> Exercise time was defined as the length of time a patient could walk on the treadmill as the grade and/or speed was increased every 3 min. The subjects' ECG and pulse oximetry were monitored by a physician. Blood pressure was also measured at each stage during the exercise and every 2 min until stable during the recovery period.

Pulmonary function tests were obtained for each patient before and after training at The Ohio State University Children's Hospital. The tests were administered by a respiratory therapist, who was blinded to the particular group the subjects were in. A spirometer and a pressure plethysmography were used for obtaining lung volumes and spirometry measurements using standard methodologies. The predicted values for spirometry were those of Knudson et al<sup>22</sup> and Weng and Levison<sup>23</sup> for lung volumes.

### Data Analysis

Descriptive statistics were computed to describe the characteristics of the subjects and their families; *t* tests and the  $\chi^2$  test (Fisher's Exact Test for expected cells <5) were used to compare the groups on demographic characteristics. For each of the major outcome variables (*ie*, inspiratory muscle strength, time on treadmill, and pulmonary functions), analysis of covariance was implemented to examine for significant differences between groups in changes from before to after values.<sup>24</sup> The initial pretreatment value of the outcome variable was used as the covariate. Using changes in pretest and posttest values and the pretest values as a covariate would adjust for possible differences between the groups in their initial pretreatment values. The normality assumptions of the residuals of these analyses were examined.

Linear regressions between the various outcome variables were used to evaluate significant correlative relationships between them. The "critical correlation coefficient," which is based on a *t* test for significance of the product-moment correlation coefficient,<sup>25</sup> was

**Table 1—Demographic Characteristics of the Children\***

Variable	Group 1 Control (n = 10)	Group 2 Experimental (n = 10)	p Value
Age, yr			
Mean	9.76	11.46	0.15
SD	2.57	2.45	
Range, yr/mo	7/5-14/3	7/9-14/11	
Gender			
M	5	6	1.00†
F	5	4	
Height, cm, mean	138	147	0.24
Weight, kg, mean	33.8	39.8	0.27
Tanner stage			
1	7	6	0.63
2	0	0	
3	2	4	
4	1	0	
NIH score, mean	86.8	87.7	0.80

\*Comparison of characteristics in patients with cystic fibrosis who trained at a constant load (control group, n = 10) and patients who trained at incremental loads (experimental group, n = 10). NIH = National Institutes of Health.

†Since expected cell counts were less than 5, the Fisher's Exact Test was used in calculating gender differences.

employed to test the significance of the regressions.

## RESULTS

### Characteristics of the Sample Populations

The control and experimental groups were compared with respect to age, sex, height, and weight, Tanner stage, and the NIH score and are reported in Table 1. There were no statistically significant differences between the two samples.

### Inspiratory Muscle Strength

Measurements of maximum inspiratory pressures before training in the two groups are illustrated in Figure 1. Note that although most children could generate pressures near 100 percent of the normal predicted value,<sup>26</sup> the ability to generate P<sub>imax</sub> decreased as a function of age (p < 0.01). A decrease in the ability to generate P<sub>imax</sub> with age would be consistent with deteriorating chest wall function as the disease process develops. The patients' NIH scores also tended to decrease with age (r = -0.4147), but this was not statistically significant. This may be due in part to the small sample size.

Figure 2 shows the effects of 10 weeks of IMT on P<sub>imax</sub>. There were no significant changes in P<sub>imax</sub> in the control group over the 10-week period. However, the experimental group significantly increased P<sub>imax</sub> an average of 13 percent (p < 0.01).

### Lung Volumes and Spirometry

Pulmonary function measurements before and after

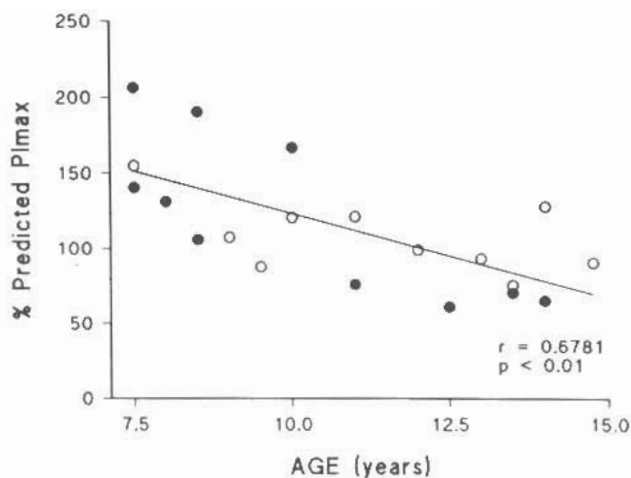


FIGURE 1. Correlation between percent predicted in P<sub>imax</sub> at pretest for both groups.

the 10-week training protocol are listed in Table 2. Although there were 10 subjects in each group, 2 tests had to be discarded. In one subject the prepulmonary function tests were inadvertently missed, and one subject did not complete the posttest. In general, the characteristics of the two groups were similar prior to training. There was little evidence of airflow obstruction, with FEV<sub>1</sub>/FVC in the controls being 84.7 ± 11 percent and in the experimental subjects, 86.2 ± 6 percent. Most patients exhibited significant elevations in RV and RV/TLC, and there was a modest degree of hyperinflation, with FRC and TLC being elevated above the predicted mean in 16 of 20 and 14 of 16 subjects in the controls and experimental subjects, respectively. We hypothesized that the degree of hyperinflation would increase as a function of age and that the degree of hyperinflation would be a reasonable

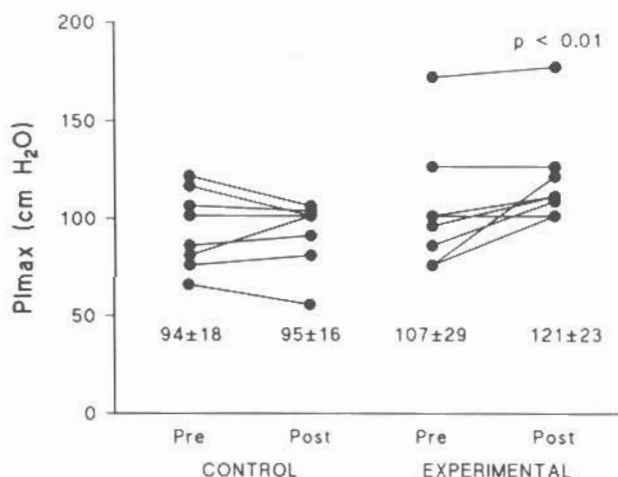


FIGURE 2. Pretest and posttest changes in maximal inspiratory muscle training for the control group and the experimental group. Pretest and posttest changes in maximal inspiratory muscle training for patients who trained at a constant load (control group, n = 10) and patients who trained at incremental loads (experimental group, n = 10).

Table 2—Pulmonary Function Tests\*

	Pretest			Posttest		p Value†
	Mean ± SD	n	% Pred	Mean ± SD	n	
VC, L						
Control	1.94 ± 0.58	9	96 ± 18	1.86 ± 0.74	9	<0.01
Experimental	1.96 ± 0.81	9	87 ± 16	2.29 ± 0.86	9	
FEV <sub>1</sub> , L						
Control	1.52 ± 0.49	9	92 ± 29	1.41 ± 0.63	8	0.10
Experimental	1.71 ± 0.81	9	89 ± 20	1.96 ± 0.86	9	
FRC, L						
Control	1.92 ± 0.68	9	138 ± 41	2.01 ± 0.90	7	0.63
Experimental	1.93 ± 0.52	9	122 ± 22	2.09 ± 0.80	9	
IC, L						
Control	1.38 ± 0.42	9	102 ± 18	1.37 ± 0.52	9	0.07
Experimental	1.48 ± 0.54	9	100 ± 41	1.70 ± 0.58	9	
RV, L						
Control	1.37 ± 0.65	9	211 ± 89	1.42 ± .79	9	0.69
Experimental	1.45 ± 0.49	9	210 ± 62	1.58 ± .69	9	
TLC, L						
Control	3.31 ± 0.94	9	114 ± 19	3.30 ± 1.17	9	<0.001
Experimental	3.41 ± 0.99	9	107 ± 17	3.86 ± 1.07	9	
RV/TLC, %						
Control	40.3 ± 10.7	9	181 ± 48	41.6 ± 13.3	9	0.26
Experimental	43.8 ± 11.4	9	196 ± 52	41.0 ± 13.1	9	
MVV, L/min						
Control	55.9 ± 25.59	9	80 ± 33	52.6 ± 23.77	9	0.12
Experimental	65.2 ± 22.65	9	81 ± 18	71.6 ± 25.33	9	
FEV <sub>1</sub> /FVC, L/min						
Control	84.7 ± 11	9	102 ± 9	80.8 ± 12	9	0.20
Experimental	86.2 ± 6	9	101 ± 16	87.4 ± 9	9	

\*Pretest and posttest comparisons in pulmonary function test results between patients who trained at a constant load (control group, n = 9) and patients who trained at incremental loads (experimental group, n = 9).

†Based on analysis of covariance on the changes from before to after (adjusting for before values) between control and experimental groups.

predictor of diminished inspiratory muscle strength. However, there were no significant relationships between the ages of the subjects and the degree of any of the pulmonary function abnormalities, nor were there any significant relationships between the percent predicted P<sub>imax</sub> and any of the percent predicted of the pulmonary function tests.

The subjects undergoing training showed clear improvements in a number of pulmonary function measurements. Vital capacity (VC) increased an average of 14 percent in the experimental group ( $p < 0.01$ ), with no similar increase in controls. Likewise, TLC increased 11.6 percent in experimental subjects, but not in controls ( $p < 0.01$ ). These increases in lung volumes appeared to be due primarily to increases in inspiratory capacity (IC), but changes in IC (mean increase = 12.9 percent) did not reach a level of statistical significance ( $p < 0.07$ ).

#### Maximal Exercise Testing/Time on Treadmill

As a group, these subjects showed normal or modest reductions in maximum exercise capacity at the pre-study evaluation. Only 9 of the 20 subjects had exercise times less than 1 SD below the average for their sex and age group.<sup>21</sup> Despite this, as shown in Figure 3,

the time the experimental subjects could continue walking on an incremental exercise test increased 10 percent in the experimental group, and this change was significantly different from the prechanges to

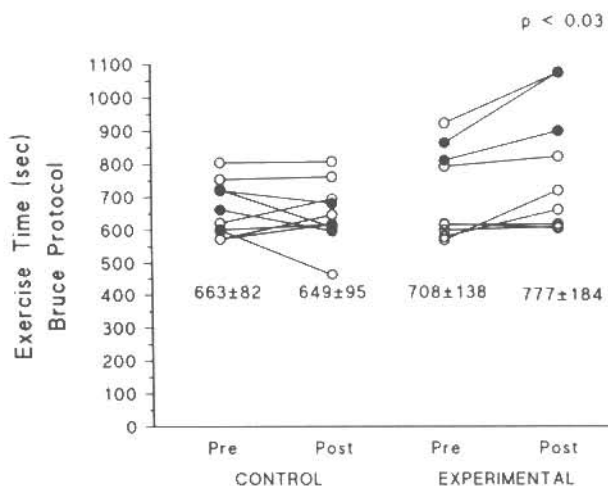


FIGURE 3. Pretest and posttest changes in exercise time for the control group and the experimental group. Pretest and posttest changes in exercise time for patients who trained at a constant load (control group, n = 10) and patients who trained at incremental loads (experimental group, n = 10).

postchanges in controls ( $p < 0.03$ ). We hypothesized that patients who showed ventilatory limitations to exercise would be most likely to demonstrate improvements in exercise capacity following IMT. Since no direct gas exchange measurements were made, it was difficult to determine whether primary ventilatory limitation was present. However, we made the assumption that patients who did not reach  $-1$  SD below the normal predicted maximum heart rate for their ages<sup>21</sup> would be most likely to exhibit ventilatory limitation to exercise. Six of the experimental subjects and eight of the controls fit this category. When tested, however, there were no significant differences between the changes observed in exercise capacity with training in the patients who had the low maximal heart rates and those who had normal predicted maximal heart rates.

We also hypothesized that the patients with the largest change in maximum voluntary ventilation (MVV), FEV<sub>1</sub>, or VC in response to training would show the largest change in exercise time. However, there were no significant relationships between changes in any of the lung volume measurements and improvement in exercise duration.

#### DISCUSSION

The primary finding of this study is that an intense IMT program can be effectively applied to a group of young patients with CF and can result in significant improvements in inspiratory muscle strength, lung volumes, and maximum exercise capacity.

The relationships between these outcome variables is still somewhat unclear in that changes in P<sub>max</sub> held very little predictive power for relative increases in lung volumes or in exercise duration. Furthermore, relative improvements in lung volumes did not predict relative improvements in exercise duration.

##### *Baseline Measurements for Both Groups of Patients*

As a group, these patients had relatively well-preserved lung function, exercise capacity, and maximum inspiratory muscle strength at the prestudy evaluation. This is no doubt due, in part, to the young age of the study population. However, the observed preservation of inspiratory muscle strength is in basic agreement with previous studies that have demonstrated it to be normal or elevated in most patients with CF.<sup>6-9</sup> We did not normalize maximum pressures to the degree of absolute lung volume at FRC because the elevation in FRC was relatively small in these patients. The drop in percent predicted P<sub>max</sub>, observed with age (Fig 1), was apparently not related to associated elevations in FRC with age in this group, because there was no trend toward age-related increases in FRC as there was for P<sub>max</sub>. We therefore conclude that the consistent decrease in percent

predicted P<sub>max</sub> with age in this group is most likely related to some poorly defined aspect of the disease process that is developing over time.

##### *Comparison of the Results of Training With Previous Studies*

Our results are in basic agreement with those of Asher et al<sup>8</sup> who used an orifice-like resistive training device in patients with CF. In both studies, a significant increase in P<sub>max</sub> was observed after training. It is unlikely that this observation reflects a learning effect. All subjects had sufficient experience with the testing methods and the training device prior to data collection to rule out the learning effect. In addition, in the present study there were no associated changes in the controls, who had similar experience performing the procedures.

Inspiratory muscle endurance was increased in the patients studied by Asher et al.<sup>8</sup> Unfortunately, whether the training protocol used in this study also improved endurance could not be evaluated because of difficulties with cooperation of the children. However, similar techniques have resulted in improvements in endurance in normal adults,<sup>27</sup> athletes,<sup>28</sup> and patients with COPD,<sup>15,16</sup> so it is likely that the endurance properties of the inspiratory muscles were increased as well as inspiratory muscle strength.

However, the results of this study differed from those of Asher et al<sup>8</sup> in that both lung volumes and maximum exercise capacity were also affected by training. One important difference between the two approaches was the length of the training program. Asher et al<sup>8</sup> trained subjects for only 4 weeks which may not have been a sufficient duration to observe the kind of influence on lung volumes and exercise capacity seen here after 10 weeks. A second difference was the training device used. The threshold loading device used in this study provided the opportunity to regulate the pressure the subjects inspired against, regardless of the pattern of breathing, and also enabled the load to be incrementally increased over the course of the 10 weeks. A particular problem with the orifice-type training device is that it is unreliable and patients learn to alter their breathing pattern as they become accommodated to the training procedures.<sup>29</sup> A third difference is the age of our study population which was approximately 6 years younger than that of Asher et al.<sup>8</sup> As will be discussed below, there may be specific age-related effects on the conditioning process that could have affected the results.

It is difficult to directly compare our results with those of Keens et al<sup>14</sup> because of the different nature of the training paradigms and the outcome variables. Our results are certainly consistent with theirs in that respiratory muscle function could be affected by training. Most of our subjects showed improvements

in ventilatory capacity (MVV) following the resistive training program, though as a group this did not reach statistical significance. Whether this also resulted in changes in sustainable ventilatory capacity or ventilatory endurance is not known.

### *Changes in Lung Volumes*

Of considerable interest were the consistent increases observed in static lung volumes in the experimental subjects. To our knowledge, lung volumes have been shown to increase in response to respiratory muscle training in only two previous studies.<sup>30,31</sup> The clearest results were reported by Fanta et al<sup>31</sup> who showed approximately 3.9 percent increases in VC in subjects who performed near-isometric inspiratory strength conditioning, close to TLC. It is interesting that using similar techniques to those applied in this study, high-intensity pressure loading with a threshold valve did not result in increases in VC in normals<sup>27</sup> or in patients with COPD.<sup>16</sup>

The above observations lead to this question: What was unique about these young patients with CF or what was unique to this training protocol that resulted in significant improvements in pulmonary function? There are several possible hypotheses.

First, these patients may be the youngest subjects in whom inspiratory muscle conditioning has been attempted, particularly at this very high level of intensity. It may be possible that the subjects' young age range, coupled with the conditioning stimulus, may have produced conditions favorable to chest wall and/or lung expansion. The chest wall is much more compliant in children between the ages of 5 to 16 years<sup>32</sup> and both the lungs and chest wall are undergoing a period of rapid change during this period. Although alveolar multiplication is generally believed to have ceased by 8 years of age, further increases in lung volume must occur primarily due to changes in alveolar dimensions.<sup>33</sup> These may be influenced greatly by changes in the shape of the chest wall via "stretch."<sup>33,34</sup> Growth hormone may be important in modulating growth of the lung and the chest wall,<sup>33</sup> and its activity in these patients during the period of conditioning was probably elevated. In addition, since total respiratory system recoil is reduced in this age range,<sup>32</sup> small changes in inspiratory muscle strength at TLC may lead to greater influences on lung volume than would be seen in adults.

A second possibility is the particular type of training protocol used for these patients. Subjects were encouraged to inspire all the way to near TLC with each breath. This may result in a length-dependent influence on inspiratory muscle strength, similar to that described by Fanta et al.<sup>31</sup> However, similar instructions have been given to adults, using similar training intensities, with no apparent increase in TLC or

VC.<sup>16,27</sup> The protocol also called for home visits to be made three times a week. Strict monitoring and encouragement might have contributed to ensuring compliance. Subjects were also eager to please and parents of this population are willing to follow a protocol that they think will contribute to their child's health. Finally, a subjective observation associated with this study was that the experimental subjects demonstrated a considerable degree of coughing and sputum mobilization during each training period, compared with control subjects. Although this effect was not quantified, it is possible that this resulted in an improvement in pulmonary toilet which may then have led to an overall improvement in pulmonary function at the time of pulmonary function testing. A rise in VC and TLC could theoretically be possible if mucous-clogged areas of the lung were opened up. Regional expansion of these previously blocked areas could presumably result in a reduction of global lung elastic recoil at TLC. Whatever the mechanism for the improved lung volumes, this phenomenon has potentially the greatest importance with respect to the long-term prognosis and health of this study population.

### *Exercise Capacity*

The standard Bruce protocol<sup>21</sup> for maximal exercise testing was used for evaluating exercise capacity. It has been suggested that children with CF cannot be tested with the usual Bruce protocol, but require an "intermittent" protocol.<sup>35</sup> This was not the case with this group of subjects, perhaps because of their relatively good functional status. In fact, during preliminary data collection, we found that the intermittent protocol was not effective in testing for maximal exercise capacity because the subjects could not reach maximum effort using this method.

The observed improvement in maximum exercise intensity in response to training was modest but of considerable significance with respect to the potential application of IMT to patients more severely ill with CF. The primary factor limiting exercise in normal children is not well understood. Although the cardiovascular system is normally considered to be primarily responsible for maximum exercise limitation in adults,<sup>36</sup> this is less clear in children. For example, the ventilatory requirement is higher in children for a given level of metabolic activity, possibly due to a higher respiratory frequency response as a fraction of VC.<sup>37</sup> Furthermore, most patients with CF did not reach within 1 SD of maximum predicted heart rate. Therefore, it is possible that ventilatory limitation may have contributed to exercise limitation in many of these patients. The improvement in exercise capacity in the subjects who underwent IMT could therefore be secondary to improved lung volumes and perhaps

the trend toward improved ventilatory capacity. However, no significant correlative relationships could be demonstrated between changes in MVV and changes in exercise capacity.

It is also possible that the high-intensity training resulted in a reduction in the sensation of dyspnea. Recent studies suggest that improvements in maximum inspiratory pressures following IMT procedures result in a reduction in the sensation of the intensity of loading in response to artificially applied resistive or elastic loads.<sup>37</sup> In addition, in patients with CF, exercise training reduces the sensation of breathlessness during exercise, at a given level of ventilation.<sup>38</sup> Therefore, the subjects undergoing IMT may have experienced a lesser sensation of respiratory effort at maximal exercise, allowing them to continue longer on the treadmill for a greater period of time.

### CONCLUSIONS

This study suggests that IMT may provide some benefit to the young CF population with respect to functional status. Whether the effects of this training protocol are carried over time and whether such procedures provide long-term benefits to this patient population are yet to be determined. Furthermore, the optimal conditioning stimulus to produce these effects needs to be determined. We do not know whether strength, endurance, or particular length-dependent or velocity-dependent training techniques<sup>39</sup> would be most beneficial to this patient group. Furthermore, we do not know whether such effects would be seen in older and more disease-impaired subjects. Nevertheless, the results are promising and may potentially be useful as an adjunct to well-established physical therapy regimens in this patient group.

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