Chest Physiotherapy in Cystic Fibrosis: Improved Tolerance With Nasal Pressure Support Ventilation

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ABSTRACT. *Objective.* **Chest physiotherapy (CPT) is an integral part of the treatment of patients with cystic fibrosis (CF). CPT imposes additional respiratory work that may carry a risk of respiratory muscle fatigue. Inspiratory pressure support ventilation (PSV) is a new mode of ventilatory assistance designed to maintain a constant preset positive airway pressure during spontaneous inspiration with the goal of decreasing the patient's inspiratory work. The aim of our study was 1) to evaluate respiratory muscle fatigue and oxygen desaturation during CPT and 2) to determine whether noninvasive PSV can relieve these potential adverse effects of CPT.**

Methods. **Sixteen CF patients in stable condition with a mean age of 13 ± 4 years participated to the study. For CPT, we used the forced expiratory technique (FET), which consisted of one or more slow active expirations starting near the total lung capacity (TLC) and ending near the residual volume. After each expiration, the child was asked to perform a slow, nonmaximal, diaphragmatic inspiration. After one to four forced breathing cycles, the child was asked to cough and to expectorate. A typical 20-minute CPT session consisted of 10 to 15 FET maneuvers separated by rest periods of 10 to 20 breathing cycles each. During the study, each patient received two CPT sessions in random order on two different days, at the same time of day, with the same physiotherapist. During one of these two sessions, PSV was provided throughout the session (PSV session) via a nasal mask using the pressure support generator ARM25 designed for acute patients (TAEMA, Antony, France). The control session was performed with no nasal mask or PSV. Both CPT sessions were performed without supplemental oxygen. Lung function and maximal inspiratory pressures (Pimax) and expiratory pressures (Pemax) were recorded before and after each CPT session.**

Results. **Mean lung function parameters were comparable before the PSV and the control sessions. Baseline pulse oximetry (Spo_2) was significantly correlated with the baseline vital capacity (% predicted) and forced expiratory volume in 1 second (FEV_1) (% predicted). PSV was associated with an increase in tidal volume (Vt) from 0.42 ± 0.01 liters to 1.0 ± 0.02 liters. Respiratory rate was significantly lower during PSV. Spo_2 between the FET maneuvers was significantly higher during PSV as compared with the control session. Spo_2 decreases after FET were significantly larger during the control session (nadir: 91.8 ± 0.7%) than during the PSV session (93.8 ± 0.6%). Maximal pressures decreased during the control session (from 71.9 ± 6.1 to 60.9 ± 5.3 cmH_2O, and from 85.3 ± 7.9 to 77.5 ± 4.8 cmH_2O, for Pimax and Pemax, respectively) and increased during the PSV session (from 71.6 ± 8.6 to 83.9 ± 8.7 cmH_2O, and from 80.4 ± 7.8 to 88.0 ± 7.4 cmH_2O, for Pimax and Pemax, respectively). The decrease in Pemax was significantly correlated with the severity of bronchial obstruction as evaluated based on baseline FEV_1 (% predicted). Forced expiratory flows did not change after either CPT session. The amount of sputum expectorated was similar for the two CPT sessions (5.3 ± 5.3 g vs 4.6 ± 4.8 g after the control and PSV session, respectively; NS). Fifteen patients felt less tired after the PSV session. Ten patients reported that expectoration was easier with PSV, whereas 4 did not note any difference; 2 patients did not expectorate. Nine patients expressed a marked and 5 a small preference for PSV, and 2 patients had no preference. The physiotherapists found it easier to perform CPT with PSV in 14 patients and did not perceive any difference in 2 patients.**

Discussion. **Our study in CF children shows that respiratory muscle performance, as evaluated based on various parameters, decreased after CPT and that significant falls in oxygen saturation occurred after the FET maneuvers despite the quiet breathing periods between each FET cycle. These unwanted effects of CPT were both reduced by noninvasive PSV delivered via a nasal mask. These data suggest that noninvasive PSV in CF patients partly compensated for the additional inspiratory overload resulting from FET, thereby decreasing the inspiratory work of breathing. This may allow the patient, assisted by a physiotherapist, to concentrate on the expiratory effort, which is the key to the efficacy of FET.**

In our study, Pimax and Pemax decreased significantly after the control session, indicating that CPT was associated with respiratory muscle fatigue. Pimax improved significantly after the PSV session. PSV delivers an unchanging level of positive pressure during spontaneous inspiration, acting as an additional external inspiratory muscle that reduces both the effort of breathing and the cost in oxygen in proportion to the level of pressure used. PSV has been shown to reduce diaphragmatic activity and to prevent diaphragmatic fatigue in chronic obstructive pulmonary disease patients. The improvement in Pimax after the PSV session in our study suggests that PSV may "rest" the inspiratory muscles during CPT. The improvement in Pemax after the PSV session could be explained by the increase in Vt during PSV. During PSV, the Vt tends to the TLC. This allows a larger amount of energy to accumulate, thereby facilitating expiration and decreasing the work of the expiratory muscles.

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The beneficial effect of PSV on Spo_2 can be explained by the large Vts, which can improve ventilation-perfusion mismatching.

Conclusions. **Our study is the first to show that PSV performed with a nasal mask during the CPT was associated with an improvement in respiratory muscle performance and with a reduction in oxygen desaturation. The improvement in patient comfort may help to improve compliance with CPT in CF patients.** *Pediatrics* **1999;103(3). URL: http://www.pediatrics.org/cgi/content/full/103/3/e32;** ***respiratory muscles, lung function, children, cystic fibrosis, oxygenation, inspiratory assistance by positive airway pressure, mucus.***

ABBREVIATIONS. CPT, chest physiotherapy; CF, cystic fibrosis; FET, forced expiratory technique; PSV, pressure support ventilation; Spo_2, pulse oximetry; FEV_1, forced expiratory volume in 1 second; TLC, total lung capacity; RR, respiratory rate; $PETCo_2$, end-tidal Co_2; $mSpo_2$, mean Spo_2; Vt, tidal volume; COPD, chronic obstructive pulmonary disease; VC, vital capacity; PEF, peak expiratory flow; FEF, forced expiratory flow; HR, heart rate; bpm, beats per minute; FRC, functional residual capacity; Pimax, maximal inspiratory pressure; Pemax, maximal expiratory pressure; ANOVA, analysis of variance; RMS, respiratory muscle strength; LS, leg strength; CPAP, continuous positive airway pressure; BiPAP, bilevel positive pressure.

Chest physiotherapy (CPT) is an integral part of the treatment of patients with cystic fibrosis (CF). The goal of CPT is to improve ventilation and mucociliary clearance through the removal of tenacious secretions obstructing the airways. The removal of these secretions may relieve atelectasis and also may prevent or slow proteolytic airway damage by clearing substances that can promote infection.[1] Although CPT is considered an effective prophylactic and therapeutic intervention in patients with CF, no studies comparing various CPT modalities are available.[2,3] One of many alternative CPT techniques is the forced expiratory technique (FET), first described by Pryor and Webbes,[4] which consists in one or two forced expirations or huffs, followed by quiet breathing. Secretions that reach the upper airways can be expectorated by one or two coughs or huffs. The FET is thought to promote secretion movement through changes in thoracic pressures and airway dynamics. The quiet breathing periods are essential to prevent bronchospasm and oxygen desaturation.[5,6] In a study by Pryor and colleagues [7] during postural drainage with assisted breathing by a physiotherapist, arterial oxygen saturation as assessed by pulse oximetry (Spo_2) did not fall during CPT when thoracic expansion exercises combined with chest clapping alternated with periods of relaxed breathing. However, this study involved discontinuous Spo_2 and therefore may have missed episodes of transient arterial oxygen desaturation.

CPT imposes additional respiratory work that may carry a risk of respiratory muscle fatigue in patients with severe respiratory disorders such as CF. Respiratory muscles have been studied extensively in stable patients with CF, but we are not aware of any studies of respiratory muscle function during CPT in CF.

Inspiratory pressure support ventilation (PSV) is a new mode of ventilatory assistance designed to maintain a constant preset positive airway pressure during spontaneous inspiration with the goal of decreasing the patient's inspiratory work. When the respiratory rate (RR), tidal volume (Vt), and inspiratory time are controlled by the patient, the work performed by the inspiratory muscles, especially the diaphragm, has been shown to be substantially reduced.[8] This method of partial mechanical ventilatory support delivered via a face mask to patients with acute exacerbations of chronic obstructive pulmonary disease (COPD) has been found to obviate the need for conventional mechanical ventilation.[9] One of the primary advantages of PSV is its good acceptability because of the fact that the frequency and duration of the inspiratory assistance are controlled by the patient rather than by the machine.[8]

We studied children with CF 1) to evaluate respiratory muscle fatigue and oxygen desaturation during CPT, and 2) to determine whether noninvasive PSV can relieve these potential adverse effects of CPT.

METHODS

The study protocol was approved by our institutional review board. Informed consent was given by all the patients and their parents.

Patients

Sixteen clinically stable patients with CF (9 girls, 7 boys) with a mean age of 13 ± 4 years (range, 6 to 18 years) were studied. The patients had to be in at baseline status, in a clinically stable phase of their disease. The mean Shwachmann score[10] was 68 ± 4 (range, 45 to 90). Nine patients were colonized with *Pseudomonas aeruginosa* and 7 with *Burkholderia cepacia*. Inhaled bronchodilators and corticosteroids were used on a long-term basis by 6 and 7 patients, respectively. Eleven patients were on RhDNase. One patient, who was on a lung transplant list, had been receiving nocturnal nasal PSV for the last 6 years. None of the patients changed their usual treatment during the study period.

The patients were divided into a "severe" group and a "moderately severe to normal" group, based on whether their forced expiratory volume in 1 second (FEV_1) was <40% or >40% of the predicted value.[11]

Chest Physiotherapy

We used the FET as described by Pryor and Webbes,[4] in which the patients increase their expiratory flow with the goal of propelling their bronchial secretions to the upper airways. In our study in children older than 6 years, the FET consisted of one or more slow active expirations starting near the total lung capacity (TLC) and ending near the residual volume. To prevent bronchial collapse by maintaining a high pressure in the airways, the expirations were done with the glottis and mouth partly closed, which produced a characteristic grunting sound. The physiotherapist assisted the expirations by applying pressure to the child's ribs. After each expiration, the child was asked to perform a slow, nonmaximal, diaphragmatic inspiration. After one to four forced breathing cycles, the child was asked to cough and to expectorate. A typical 20-minute CPT session consisted of 10 to 15 FET maneuvers, separated by rest periods of 10 to 20 breathing cycles each. All the patients were familiar with this CPT technique and used it at home once or twice a day.

During the study, each patient received two CPT sessions in random order on two different days, at the same time of day, with the same physiotherapist. During one of these two sessions, PSV was provided throughout the session (PSV session) via a nasal mask using the pressure support generator ARM25 designed for acute patients (TAEMA, Antony, France). This inspiratory-assistance device functions on three principles. First, during the patient's spontaneous inspiration, positive pressure is generated in the circuit by a jet of compressed air delivered through a tube open to the atmosphere. As a result, the patient inspires in a

pressurized circuit. The airway pressure remains approximately constant over a wide range of inspiratory flows (for example, pressure decreases only from 15 to 12 cmH_2O when the inspiratory flow increases from 0 to 1.5 L per second). Second, the beginning and the end of the patient's inspiration are sensed by a hot wire-calibrated pneumotachograph placed on the inspiratory limb of the circuit. An inspiratory flow of 2 $l.min^{-1}$ triggers the positive-pressure assistance, which is driven by an electronic valve. The pressure assistance stops when the inspired flow falls below a preset threshold. Third, the expiration is not assisted and takes place through a low-resistance expiratory line that is separated from the inspiratory line by a pneumatic valve open to the atmosphere.[9] These principles were explained to the patients. Before the PSV CPT session, a nasal mask (Sullivan, Resmed Ltd, North Ryde, Australia, or Respironics Inc, Murrysville, PA) was applied, and pressure support was increased gradually from 6 cmH_2O to 12 cmH_2O, which was the level of pressure support associated with maximal patient comfort. The PSV expiratory threshold was set at 10 $l.min^{-1}$. The patients wore the nasal mask throughout the session and were therefore able to perform the FET through the mouth. PSV was applied during the inspiratory part of the FET maneuvers and during the resting periods between the FET maneuvers. Expectoration was done without removing the mask.

The control session was done with no nasal mask or PSV. As a result, Vt could not be measured during this session.

Both CPT sessions were performed without supplemental oxygen.

After each CPT session, the patients were asked to report their subjective impressions (fatigue, ease of expectoration, and preference). The physiotherapists also gave their subjective impression regarding the effect of nasal PSV on CPT and were asked specifically to compare the CPT sessions with and without nasal PSV in each patient, using one of three ratings, namely, "worse (= −1)", "the same (= 0)", or "better (= +1)". These subjective impressions were evaluated by different, independent persons who were not involved in the study and who were not aware of the CPT regimen. A marked preference corresponded to a score of 2 or 3, a small preference to a score of 1.

Secretions expectorated during and after each CPT session were collected and weighed.

Lung Function Testing

Lung function testing was conducted using a spirometer (Morgan spirometer, PK Morgan Ltd, Chatham, Kent, UK). Vital capacity (VC); forced vital capacity (FVC); FEV_1; peak expiratory flow (PEF); and forced expiratory flows at 50%, 25%, and 25% to 75% of FVC (FEF_{50}, FEF_{25}, and $FEF_{25–75}$, respectively) were recorded before and after each CPT session. The best of three reproducible flow curves was analyzed.[11]

Airway resistance by the flow interruption technique (Spirograph Booster, EMO International, La Rochelle, France) was recorded before and after each CPT session. Values were expressed as absolute values and as a percentage of the predicted value for height and sex.[12]

Maximal inspiratory pressures (Pimax) were measured at the end-expiratory volume (FRC), and expiratory pressures (Pemax) at TLC after maximal inspiration. Pressures were generated at the mouth against a closed tap, with the nose occluded by a nose clip and the subject seated. A small leak in the circuit was used during both maneuvers. Pressures had to be maintained for at least 1 second. At least three Pimax and Pemax values, separated by at least 1 minute, were obtained before and after each CPT session. The mean of three maximal efforts was calculated.[13,14]

TABLE 1. Lung Function Parameters Before the Two Chest Physiotherapy Sessions

	Control Session	PSV Session	*P*
FVC (% predicted value)	60.3 ± 5.8	61.5 ± 5.7	NS
FEV_1 (% predicted value)	51.9 ± 6.5	51.3 ± 6.4	NS
FEF_{50} (% predicted value)	47.7 ± 8.3	44.6 ± 8.0	NS
FEF_{25} (% predicted value)	38.7 ± 6.7	34.6 ± 6.7	NS
$FEF_{25–75}$ (% predicted value)	52.4 ± 8.3	45.7 ± 1.5	NS
PEF (% predicted value)	60.1 ± 7.0	54.1 ± 6.3	NS
Airway resistance (% predicted value)	180 ± 13	184 ± 13	NS
PEmax (cmH_2O)	85.3 ± 7.9	80.4 ± 6.8	NS
PImax (cmH_2O)	71.9 ± 6.1	71.6 ± 8.6	NS

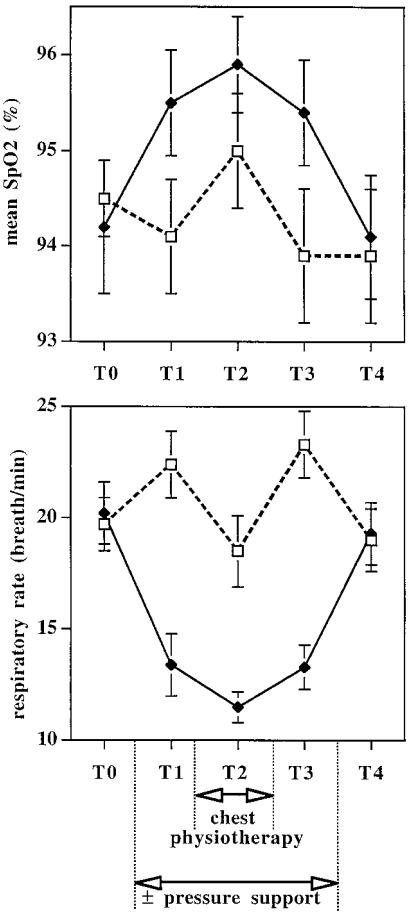

Fig 1. Changes in Spo_2 during the various study periods. T0 indicates a 2-minute period of quiet breathing; T1, 5-minute period of quiet breathing with or without nasal PSV; T2, 20-minute chest physiotherapy session with or without nasal PSV; T3 and T4 periods after physiotherapy, same conditions as for T1 and T0, respectively. For T0, T1, T3 and T4, Spo_2 values are the mean values determined over the entire period, whereas for T2, Spo_2 is the mean value measured during the quiet breathing periods between the FET maneuvers. During the control session (dashed line), Spo_2 showed a slight but significant increase during T2 compared with the other periods (ANOVA, $P < .001$). Major improvements were observed during the PSV session (solid line) (ANOVA, $P < .0001$). The periods with application of PSV were characterized by higher Spo_2 values compared with T0 and T4 of the same session, and also to all the Spo_2 values during the control session ($P < .0004$). RR decreased during the PSV session (<.0001), whereas no changes were seen during the control session.

Study Protocol

After lung function testing as described above, the patients were connected to a pneumotachograph and asked to breath spontaneously for 2 minutes (T0) to obtain the basal Vt. Respiratory airflow was measured using a pneumotachometer (Fleisch #2, Lausanne, Switzerland) connected to a pressure transducer (MP45 model, Validyne ± 2 cmH_2O, Northridge, CA) and integrated to yield Vt. Spo_2, RR, heart rate (HR), and end-tidal Co_2 ($PetCo_2$) (Ultracap, Nellcor Puritan-Bennett, Courtaboeuf, France) also were recorded during both the control and the PSV sessions. All the physiologic parameters, Sao_2, RR, HR, and $PetCo_2$, were sampled for analysis using an analogic/numeric acquisition system (MP100, Biopac Systems, Goletta, CA).

During the next 5 minutes (T1), the patient breathed either freely (no mask) or with a nasal mask and PSV. Spo_2, RR, HR,

$P_{ET}CO_2$ and PSV flow were recorded continuously during the PSV session. The T1 period allowed us to observe the effects of nasal PSV on the breathing pattern (however, Vt was measured only during the PSV session).

During the 20-minute CPT session (T2), the same parameters were recorded.

The T3 and T4 periods after the CPT sessions were similar to the T1 and T0 periods, respectively.

Analysis of Spo_2 During Chest Physiotherapy

Spo_2 during CPT (T2) involved measurement of the following parameters: 1) mean Spo_2 ($mSpo_2$) describes the oxygenation during the quiet breathing periods between the FET maneuvers; and 2) the fall in Spo_2 during the maneuvers was expressed in 3 different ways: a) the largest fall expressed in the absolute value of Spo_2 ($nadirSpo_2$); b) the largest fall expressed as a difference with the Spo_2 just before the maneuver: (ΔSpo_2 max); this parameter was computed because the baseline values of Spo_2 were different between the 2 groups); and c) the mean of ΔSpo_2 max during the whole CPT period (ΔSpo_2 mean).

Statistical Analysis

Results are expressed as mean values ± SEM. Parameters measured before and after each PSV or control CPT session were compared using a paired *t* test, with each child serving as his or her own control. Student's *t* test was used to compare the two disease severity groups. The time course of the parameters during the various periods of the PSV and control sessions was evaluated using one-way analysis of variance (ANOVA) with repeated measurements. Comparisons of these parameters during the various periods of the PSV and control sessions were performed using two-way ANOVA. Correlations between variables were analyzed using least-square linear regression technique.

The level of significance was set at 5%.

RESULTS

Functional Status at Baseline

All the patients completed the protocol. Mean lung function parameters were comparable before the PSV and the control sessions (Table 1). Figure 1 shows that Spo_2 was slightly lower before the PSV session than before the control session (94.2 ± 0.7% vs 94.5 ± 0.5%; $P < .05$). Spo_2 was significantly lower in the "severe" group ($n = 7$) than in the "moderately severe to normal" group ($n = 9$) (93.9 ± 0.5% vs 96.7 ± 0.3%; $P < .005$). Spo_2 was significantly correlated with the baseline VC (% predicted; $P = .016$) and FEV_1 (% predicted; $P < .008$).

The nutritional status of the patients was correct with no significant difference of the *z* score[15] between the two groups (−0.22 vs −0.63 for the "severe" and the "moderately severe to normal" groups, respectively.

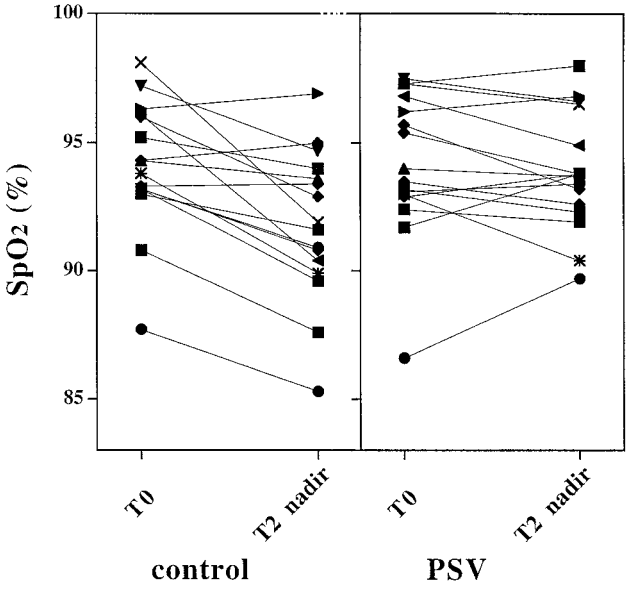

Fig 2. Time course of Spo_2 during chest physiotherapy. Spo_2 was evaluated as the $mSpo_2$ value during T0 and as the lowest Spo_2 value after the FET maneuvers during T2. During the control session, falls in Spo_2 occurred after the FET maneuvers in 13 patients. These episodes of desaturation were improved by PSV: Spo_2 decreased in only 9 patients, and the magnitude of the decrease was consistently smaller than during the control session (91.8 ± 0.7% vs 93.8 ± 0.6 for the $nadirSpo_2$ during the control and PSV sessions, respectively; $P < .0007$).

Changes in RR, $P_{ET}CO_2$, and HR During Chest Physiotherapy

$P_{ET}CO_2$, RR, and HR remained stable throughout each of the four periods (including T2) of the PSV and control sessions.

HR during T2 was higher than during the other periods (T0, T1, T3, T4) during both the PSV and the control sessions. The HR increase from T0 to T2 was from 88 ± 19 beats per minute (bpm) to 96 ± 5 bpm during the control session, and from 92 ± 4 bpm to 95 ± 5 bpm during the PSV session. $P_{ET}CO_2$ declined somewhat from T0 to T2 (from 33 ± 1 mm Hg to 30 ± 1 mm Hg during the control session, and from 34 ± 1 mm Hg to 32 ± 1 mm Hg during the PSV session). PSV was associated with an increase in Vt from 0.42 ± 0.01 liters during T0 to 1.0 ± 0.02 liters during T1 ($P < .0001$).

Compared with the control session, RR was signif-

TABLE 2. Efficacy of PSV in Limiting Falls in Spo_2 After the FET Maneuvers During Chest Physiotherapy (T2 Periods)

		Control Session	PSV Session	*P*
$mSpo_2$	All subjects	94.8 ± 0.6	95.9 ± 0.5	<.0004
	$FEVI_1$<40%	93.0 ± 0.6	94.0 ± 0.5	<.05
	FEV_1>40%	96.1 ± 0.4	96.7 ± 0.3	<.003
$nadirSpo_2$	All subjects	91.8 ± 0.7	93.8 ±0.6	<.0007
	$FEV_1$40%	90.2 ± 0.8	92.0 ± 0.4	NS
	FEV_1>40%	93.0 ± 0.5	95.3 ± 0.4	.0006
ΔSpo_2 max	All subjects	−3.6 ± 0.7	−2.0 ± 0.3	<0.04
	FEV_1<40%	−4.0 ± 0.9	−2.1 ± 0.3	NS
	FEV_1>40%	−3.3 ± 0.5	−1.9 ± 0.4	NS
ΔSpo_2 mean	All subjects	−1.2 ± 0.3	−0.2 ± 0.2	<.006
	FEV_1<40%	−1.3 ± 0.3	−0.0 ± 0.2	<.02
	FEV_1>40%	−1.1 ± 0.3	−0.3 ± 0.2	NS

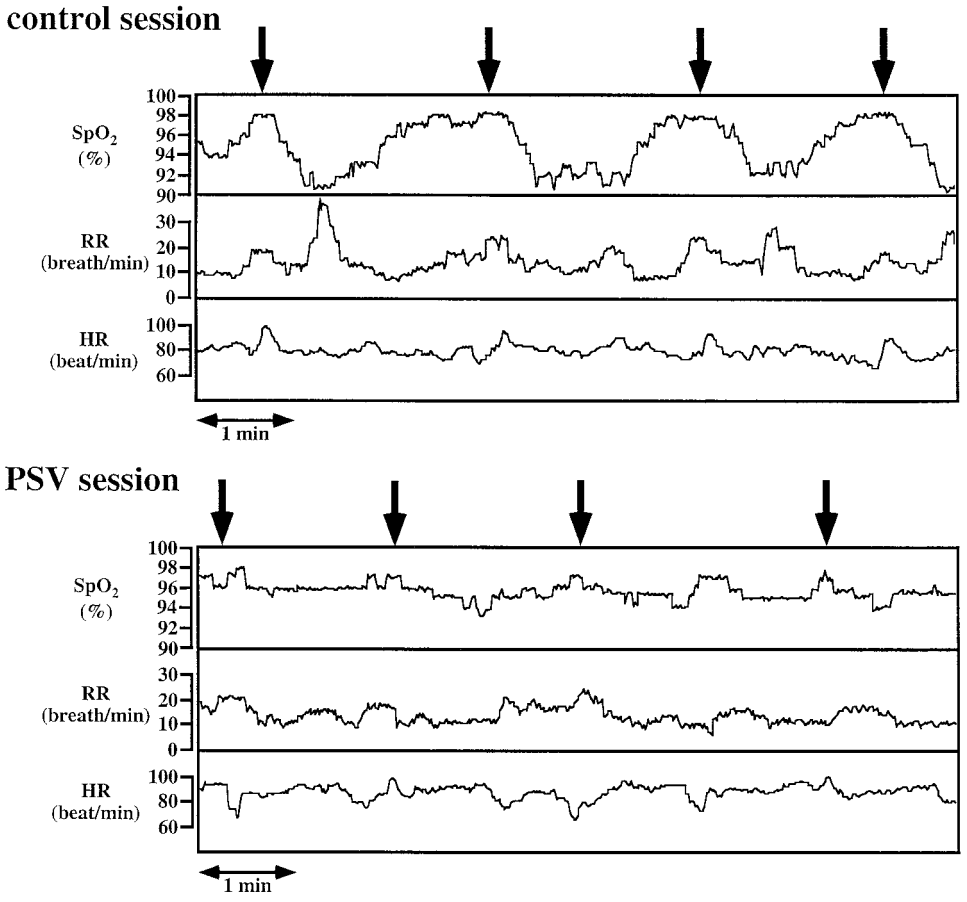

Fig 3. Time course of Spo_2, RR, and flow during the PSV session in a patient. Spo_2 fell as low as 90% during the control session, but remained between 94% and 98% during the PSV session. RR also was lower during the PSV session compared with the control session. HR did not differ between the two sessions. The arrows represent the FET maneuvers.

TABLE 3. Time Course of Lung Function Parameters After the Control and PSV Chest Physiotherapy Sessions

	Control Session			PSV Session		
	Before	After	*P*	Before	After	*P*
FVC (% predicted value)	60.3 ± 5.8	60.7 ± 5.0	NS	61.5 ± 5.7	61.1 ± 5.0	NS
FEV_1 (% predicted value)	51.9 ± 6.5	51.3 ± 6.2	NS	51.3 ± 6.4	50.4 ± 5.7	NS
FEF_{50} (% predicted value)	47.7 ± 8.3	44.6 ± 8.0	NS	40.7 ± 6.5	40.4 ± 6.6	NS
FEF_{25} (% predicted value)	38.7 ± 6.7	36.9 ± 8.3	NS	34.6 ± 6.8	30.6 ± 5.7	NS
$FEF_{25–75}$ (% predicted value)	52.4 ± 8.3	48.1 ± 9.1	NS	45.7 ± 7.4	42.1 ± 6.6	NS
PEF (% predicted value)	60.1 ± 7.0	54.5 ± 6.7	<.02	54.1 ± 6.3	52.5 ± 5.7	NS
Airway resistance (% predicted value)	180 ± 13	168 ± 9	NS	184 ± 13	159 ± 7	<.04

icantly lower during PSV (T1 to T3 of the PSV session) ($P < .0001$) (Fig 1).

Changes in Spo_2 Parameters During Chest Physiotherapy

CPT during the control session (T2) was responsible for a significant ($P < .001$) increase in $mSpo_2$ (measured during the quiet breathing periods between the FET maneuvers) versus periods T0 and T4 (Fig 1). During the PSV session, $mSpo_2$ was significantly higher during the three periods with PSV (T1 to T3) than at baseline ($P < .0001$). $mSpo_2$ was significantly higher during PSV compared with the control session ($P < .0004$)

We also examined the lowest Spo_2 values after FET. Marked decreases in Spo_2 occurred after FET during the control session. With PSV, these decreases were noticeably smaller (Table 2; Figs 2, 3).

The improvement provided by PSV in $mSpo_2$ during the intervals between the FET maneuvers was similar in the "severe" group and the "moderately severe to normal" group. As a result, $mSpo_2$ and $nadirSpo_2$ were lower in the "severe" group than in the "moderately severe to normal" group during both the control and the PSV sessions (Table 2).

Lung Function Parameters

The only significant change in spirometry parameters was a small decrease in PEF after the control session. A small but significant decrease in airway resistance occurred after the PSV session (Table 3).

Both Pimax and Pemax decreased significantly af-

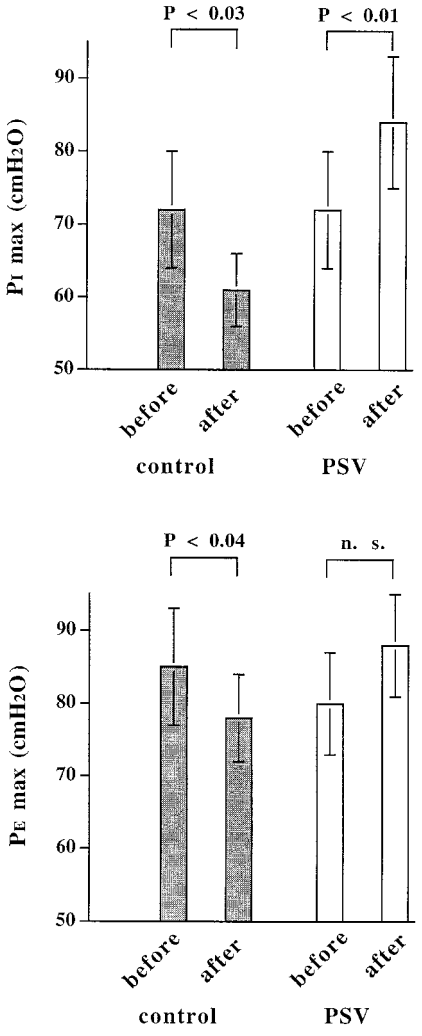

Fig 4. Changes in PImax and PEmax during chest physiotherapy. Both PImax and PEmax decreased significantly after the control chest physiotherapy session. After the PSV session, both PImax and PEmax increased, but the difference was significant only for PImax.

ter CPT during the control session (Fig 4). The decrease in PEmax was significantly correlated with the severity of bronchial obstruction as evaluated based on baseline FEV_1 (% predicted) ($P = .03$).

It is worth noting that during the PSV session, a significant increase in PImax versus baseline was seen in every patient, contrasting with the decrease seen during the control session. Both the decrease in PImax during the control session and the increase during the PSV session were highly significant (Fig 4). A nonsignificant increase in PEmax was seen during the PSV session.

The amount of sputum expectorated was similar for the two CPT sessions (5.3 ± 5.3 g vs 4.6 ± 4.8 g after the control and PSV sessions, respectively; NS).

Subjective Impression

Fifteen patients felt less tired after the PSV session. Ten patients reported that expectoration was easier with PSV, whereas 4 did not note any difference; 2 patients did not expectorate. Nine patients expressed a marked (score +3 or +2) and 5 a small preference (score +1) for PSV, and 2 patients had no preference (score 0).

The physiotherapists found it easier to perform CPT with PSV in 14 patients and did not perceive any difference in 2 patients. Headache did not occur after the PSV session in the 2 children who complained of this symptom after the control session. A 17-year-old girl told us that the PSV session made her realize for the first time how large an effort spontaneous breathing was for her.

DISCUSSION

Our study in children with CF found 1) that respiratory muscle performance, as evaluated based on various parameters, decreased after CPT, and 2) that significant falls in oxygen saturation occurred after the FET maneuvers despite the quiet breathing periods between each FET cycle. These unwanted effects of CPT both were reduced by noninvasive PSV delivered via a nasal mask. These data suggest that noninvasive PSV in patients with CF compensated partly for the additional inspiratory overload resulting from FET, thereby decreasing the inspiratory work of breathing. This may allow the patient, assisted by a physiotherapist, to concentrate on the expiratory effort, which is the key to the efficacy of FET.

Respiratory Muscles

Maximal respiratory pressures have been found to remain unchanged in stable adults with CF,[15–22] probably as a result of a selective training effect attributable to chronic lung disease.[18,20] Respiratory and peripheral muscle function was evaluated by Lands and co-workers[16] in 14 stable patients with CF and 16 healthy controls. The patients with CF had no loss in respiratory muscle strength (RMS) as measured using static maximal pressures performed at FRC, but had decreased leg strength (LS). In the group with CF, expiratory RMS and LS correlated with lean body mass and with each other, whereas inspiratory RMS was independent of lean body mass and LS, consistent with a selective training effect attributable to chronic lung disease.[16]

A recent study investigated inspiratory muscle function in stable children with CF (mean age 11 ± 2 years) versus healthy control subjects.[17] Lung function in patients with CF was moderately impaired, with a mean FEV_1 of 81 ± 16% of predicted value and a FRC of 133 ± 19% of predicted value. Inspiratory muscle function was impaired in the patients with CF despite a good nutritional status. It is known that children with COPD have a lower than predicted PImax value at FRC,[18] but that this decrease disappears after lung volume correction. Hyperinflation may be a major determinant of inspiratory muscle weakness because it puts the inspiratory muscles at a less favorable location of their length–tension curve and causes them to undergo geometrical alterations.[19]

The PImax of our patients was significantly decreased compared with predicted values[13,14] and was lower than the values reported by Hayot and associates.[17] We did not investigate lung volumes, but the

mean FEV_1 was lower in our group (52 ± 7% of predicted value) than in other studies, suggesting that hyperinflation was present in most of our patients.

Controversy exists concerning the role of malnutrition in respiratory muscle performance. Malnutrition was associated with impaired inspiratory muscle performance in most studies.[20,21] However, in one study of patients with CF and asthma, nutritional status had no influence on RMS, and this finding was ascribed to a training effect of the increased work of breathing.[22] We did not observe any relationships linking *z* score, lung function parameters, Pimax, and Pemax. This may be attributable to the relatively good nutritional status and to the young age of our patients.

Pemax has been studied less extensively than Pimax.[16,20,22,23] In one study, Pemax was similar in adult patients with CF and in healthy subjects despite significant malnutrition in some of the patients with CF.[22] In another study, the reduction in Pemax was similar to that in Pimax in subjects with malnutrition and/or hyperinflation.[20] Lands and co-workers[16] found that Pemax correlated with LS and lean body mass, suggesting that the inspiratory muscle training effect attributable to chronic lung disease may not apply to expiratory muscles. Expiratory muscle performance has been shown to be related to cough efficiency.[24,25]

Our CPT sessions can be likened to endurance tests, with repeated expiratory and inspiratory efforts during 20 minutes in children with moderate to severe lung impairment. In a pediatric study, the fraction of Pimax developed by the inspiratory muscles for breathing at rest was significantly increased, suggesting that even a tiny increase in breathing load may expose children with COPD to respiratory muscle fatigue.[18] Two previous studies have investigated inspiratory muscle endurance during inspiratory resistance breathing or repeated static maximal efforts in adults with CF.[16,26] No significant differences with respect to respiratory muscle fatigue were observed between the control subjects and the patients with CF. Compared with these two studies, our patients were younger, performed longer CPT sessions, and had more severe bronchial obstruction, which may explain the discrepancies between those results and our findings. Furthermore, the mean FEV_1 of the patients with CF in the study by Lands and coworkers was 72.5 ± 24.8% of predicted value.

Pimax improved significantly after the PSV session. PSV delivers an unchanging level of positive pressure during spontaneous inspiration, acting as an additional external inspiratory muscle that reduces both the effort of breathing and the cost in oxygen in proportion to the level of pressure used.[8] PSV has been shown to reduce diaphragmatic activity and to prevent diaphragmatic fatigue in patients with COPD.[8] The improvement in Pimax after the PSV session in our study suggests that PSV may "rest" the inspiratory muscles during CPT. In one study,[27] long-term nasal ventilation in 4 adults with CF significantly improved both Pimax and Pemax. The decrease in Pimax provided by PSV in our study also may have been in part attributable to a decrease in FRC. PSV reduces intrinsic positive end-expiratory pressure[28] as a result of a decrease in RR attributable to an increase in the duration of expiration. A decrease in FRC allows the subject to develop a higher Pimax. We did not measure lung volumes in our study, but the significant decrease in airway resistance after the PSV session supports this hypothesis.

FEV_1% predicted value was significantly correlated with the time course of Pemax. The fall in Pemax after the control session was greatest in the patients with the lowest FEV_1% predicted values. Expiratory effort increases with the severity of bronchial obstruction. The improvement in Pemax after the PSV session could be explained by the increase in Vt during PSV. During PSV, the Vt tends to the TLC. This allows to accumulate a larger amount of energy and thereby to facilitate expiration and to decrease the work of the expiratory muscles.

No significant improvements in lung function parameters were observed after either of the two CPT sessions. This finding is in agreement with recently published metaanalyses and review articles concluding that no CPT modality has been proved to be superior to another and that the short-term benefits of various CPT techniques on lung function parameters are moderate and similar in magnitude.[2,3]

Spo_2

Significant falls in Spo_2 occurred after the FET maneuvers in our study. Desaturation has been described during CPT.[5,29] It has been suggested that desaturation can be prevented by allowing sufficiently long periods of quiet breathing between the FET maneuvers.[7] In our study, desaturation occurred immediately after the FET maneuvers during the control session, consistent with a role of bronchial collapse and prolonged expiration. Bronchial collapse is frequent in CF and increases ventilation–perfusion mismatching. Mean nadirSpo_2 was as low as 91.8% in our overall study population and 90.2% in our "severe" group, and some patients had values as low as 84%. It is worth noting that in the course of the 20-minute CPT session, a mean of 13 FET maneuvers were performed, and that each was followed by desaturation.

PSV was associated with less desaturation, even in the "severe" group, in which the improvement was of the same magnitude as in the "moderately severe to normal" group. The beneficial effect of PSV can be explained by the large Vts, which can reduce bronchial collapse and improve ventilation–perfusion mismatching.

The optimal level of PSV was determined empirically based on both the subjective impression of comfort reported by each patient and on the change in the breathing pattern with a reduction in RR and an increase in Vt.[8,9] All the patients completed the study, and even the youngest adapted to nasal PSV after no more than a few breaths. Even the children with nearly normal lung function reported greater comfort and less fatigue with PSV.

Sputum Expectoration and Subjective Impression

The goal of use in our study of a noninvasive mechanical ventilation device designed for intensive care was to improve the tolerance of CPT in terms of respiratory muscle fatigue and oxygenation, rather than to improve lung function parameters or sputum expectoration. No increase in the amount of sputum was obtained in previous studies using other types of mechanical devices. Mucus transport was not influenced by positive end-expiratory pressure during CPT.[30,31] In our study, the volume of wet sputum was similar during the control and PSV sessions, demonstrating that nasal PSV during CPT did not impair sputum expectoration. The patients were able to expectorate normally despite the nasal mask because each inspiration was triggered by the patient and because a sufficiently long period for expiration was allowed.

Nasal mechanical ventilation has been used previously in patients with CF in two different conditions. In individual patients with an end-stage lung disease, nasal mechanical ventilation has been used as a "bridge to transplantation."[32] More recently, the efficacy of nasal continuous positive pressure (CPAP) or bilevel positive pressure (BiPAP) on sleep disturbance or oxyhemoglobin desaturation was analyzed in 7 patients with CF.[33] CPAP resulted in a significant improvement of Spo_2 and a reduction in sleep disturbance. Another study compared the effects of BiPAP to those of oxygen in 6 patients with CF.[34] BiPAP improved sleep-related hypoxemia and hypercapnia. But these studies compared only two nights. According to our results, we could propose nasal PSV during CPT as a third potential indication of nasal mechanical ventilation in patients with CF.

CPT is considered effective as a long-term prophylactic and therapeutic intervention. Compliance may be a major determinant of the efficacy of CPT in CF. The increase in comfort and reduced impression of fatigue provided by PSV conceivably may translate into improved compliance with CPT in patients with CF.

In conclusion, we observed a significant decrease in respiratory muscle performance after CPT in 16 children with CF with moderately severe to severe lung function impairment. During the 20-minute CPT session, significant falls in Spo_2 were observed after the FET maneuvers, even in the patients with moderately severe impairment. Our study is the first to show that PSV performed with a nasal mask during the CPT was associated with an improvement in respiratory muscle performance and with a reduction in oxygen desaturation.

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