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Effective Home-Based Pulmonary Rehabilitation in Patients with Restrictive Lung Diseases

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Patients with chronic obstructive pulmonary disease (COPD) are commonly referred for pulmonary rehabilitation (PR), but the use of PR is not common for patients with restrictive lung disease, neuromuscular diseases, and those who have sustained a severe respiratory illness or undergone thoracic surgery. We investigated the effects of PR in patients with restrictive lung diseases in comparison with COPD patients using a home-based setting. Twenty-six restrictive lung diseases patients and 40 COPD patients who had a Medical Research Council (MRC) dyspnea score ≥ 2, a clinically stable condition, and who had completed a 6-month PR program, were enrolled in the present study. The definition of restrictive lung disease was a forced vital capacity (FVC) of ≤ 80% of the predicted value with a forced effort volume in one second/FVC of > 70%. Our PR consisted of breathing retraining, exercise training, respiratory muscle stretching calisthenics, level walking, inspiratory and expiratory muscle exercises, and a monthly education program. Patients were strongly instructed to practice this program daily at home, and were supervised by a respiratory therapist every 2 weeks in our hospital. Patients with restrictive lung diseases showed the significant increases in inspiratory and expiratory muscle forces, the 6-minute walking distance, the Chronic Respiratory Disease Questionnaire and the Short-Form 36, and decreased MRC scores after 6 months. In conclusion, our home-based PR improves respiratory muscle forces, exercise tolerance, health-related quality of life, and the perception of dyspnea in patients with restrictive lung disease to the same extent as in COPD patients. ———— Pulmonary rehabilitation; restrictive lung disease; exercise tolerance; health-related quality of life.

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Pulmonary rehabilitation (PR) is one of the most important non-pharmacological treatments for patients with chronic obstructive pulmonary disease (COPD) (GOLD Workshop Report 2006). For example, PR improves exercise tolerance, health-related quality of life (HRQOL), and perception of dyspnea. PR is also recommended for some patients with chronic respiratory diseases other than COPD, but there are only limited supporting scientific data (Ries et al. 2007). One study showed that 3 months sub-maximal physical exercise in an outpatient basis for asthmatic patients improved fitness and cardiorespiratory performance (Cochrane and Clark 1990), while another demonstrated that the 6-minute walking distance (6MWD) and peak cycling load increased significantly in lung cancer patients following an intensive 8 week multidisciplinary inpatient PR (Spruit et al. 2006). In patients with cystic fibrosis, a training effect of 12 months of individualized unsupervised exercise was also demonstrated (Moorcroft et al. 2004). Some studies compared the effectiveness of PR in non-COPD patients to that of COPD patients, and concluded that the effectiveness of PR was similar (Foster and Thomas 1990; Ando et al. 2003; Ferreira et al. 2006). An improvement in the 6MWD was shown following a 4 week intensive inpatient program (Foster and Thomas 1990), while a 9 week low-intensity outpatient PR improved dyspnea score, activities of daily living, and 6MWD in patients with sequelae tuberculosis (Ando et al. 2003). Ferreira et al. (2006) also showed that an 8 week outpatient PR improved exercise tolerance and HRQOL.

However, the use of PR is not common for patients with restrictive lung disease, neuromuscular diseases, and those who have sustained a severe respiratory illness or undergone thoracic surgery (Crouch and MacIntyre 1998). Furthermore, it is unknown whether home-based PR for non-COPD patients is effective. Thus, the aim of the present study was to investigate the effects of PR in patients with restrictive lung diseases compared with COPD patients using a home-based setting.

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Materials and Methods

Forty-one patients who were diagnosed with restrictive lung disease between June 1998 and August 2004 were enrolled in the present study. The definition of restrictive lung disease was a forced vital capacity (FVC) of ≤ 80% of the predicted value with a forced effort volume in one second (FEV_1)/FVC of > 70%. Inclusion criteria were a Medical Research Council (MRC) dyspnea score (Fletcher 1960) ≥ 2, a clinically stable condition, and the absence of significant associated medical problems that might interfere with the patients abilities to undergo PR. Patients who could not visit our hospital every 2 weeks were excluded. Five patients refused to participate and 10 dropped out within 6 months. Therefore, 26 patients who gave informed consent and completed our PR program for 6 months were used for analysis; patients included 18 with sequelae tuberculosis, 4 with spinal caries, and 4 with other diseases. Forty FEV_1-matched COPD patients who gave informed consent and completed the same PR program for 6 months between June 1998 and September 2002 were selected as the control group. The Ethics Committee approval was not obtained because the Committee at the institution within which the work was undertaken did not exist when this study was started. Nevertheless, this research was conducted according to the highest ethical standards and conformed to the provisions of the Declaration of Helsinki, 1995.

The PR used in the present study was a multidisciplinary home-based program. Breathing retraining consisted of pursed-lip breathing, diaphragmatic breathing, and slow-deep breathing, both in the supine and sitting positions. Exercise training included upper and lower limb exercises, respiratory muscle stretching calisthenics (Homma 1999), level walking for at least 15 min, and inspiratory and expiratory muscle exercises using Threshold® (HealthScan Products Inc., Cedar Grove, NJ) set at a training intensity of 20-30% of the maximal inspiratory (PImax) and expiratory (PEmax) muscle forces. Patients also underwent a monthly 45 min education program including lectures and discussions on respiratory disease, control of dyspnea, medication and equipment use, nutrition, stress management, relaxation techniques, home exercises, and the benefits of PR. Patients were strongly instructed to practice this program daily at home and were supervised by a respiratory therapist every 2 weeks in our hospital. Approximately 75% of the patients were able to perform our daily program at home. The achievements of our home program have been reported (Miura et al. 2001). A nurse periodically visited each patient at home and provided information on the role of the PR program. The overall training intensity was set at a dyspnea rating scale of 3, which corresponds to approximately 50% of the maximum oxygen consumption (Horowitz et al. 1996).

Age, height, weight, body mass index (BMI), and blood gas data were evaluated at baseline. FVC and FEV_1 were measured using a lung function analyzer (CHESTAC-25 PART II EX; CHEST MI Inc., Tokyo, Japan). PImax and PEmax were evaluated following the Black and Hyatt technique (Black and Hyatt 1969), and were determined as the pressure that could be sustained for more than 3 seconds using VITALOPOWER KH-101 (CHEST MI Inc., Tokyo, Japan). Exercise tolerance was evaluated using 6MWD on a flat circuit. 6MWD was interrupted as a result of dyspnea or leg fatigue, ≤ 85% of saturation, or threatening symptoms such as angina pectoris. The Borg scale (Borg 1982) was measured after the 6MWD test. The HRQOL was assessed using the Chronic Respiratory Disease Questionnaire (CRQ) (Guyatt et al. 1987) and the Short-Form 36 (SF-36) (Fukuhara et al. 1998a; 1998b). A high score indicated good HRQOL in both CRQ and SF-36. Weight, BMI, FVC, FEV_1, PImax, PEmax, MRC, 6MWD, CRQ, and SF-36 were also evaluated at 6 months. Previous answers in the CRQ were reported to the patients as a reminder to make the questionnaire more sensitive to alterations (Guyatt et al. 1989).

Statistical analysis

Unless otherwise stated, values are expressed as mean ± standard deviation. Mann-Whitney U test was conducted to compare the variables at baseline between both groups. Wilcoxon signed rank test was applied to assess differences at the baseline and 6 months evaluation. Repeated-measure ANOVA was conducted to compare the effectiveness of PR between both groups. All analyses were performed using the StatView 5.0 for Windows statistical package (SAS Institute Inc., Cary, NC). P values less than 0.05 were considered significant.

Results

Baseline characteristics of both groups can be seen in Table 1. At baseline, FEV_1/FVC was significantly higher, FVC (% predicted) was significantly lower, and $PaCO_2$ was significantly higher in the restrictive lung diseases group compared with the COPD group. Age, height, weight, BMI, FEV_1, PImax, PEmax, PaO_2, 6MWD, and Borg scale were not different between the groups.

The MRC dyspnea scores at baseline in patients with restrictive lung diseases were grade 2 in 5 patients, grade 3 in 18 patients, and grade 4 in 3 patients, while those in COPD patients are grade 2 in 15 patients, grade 3 in 22 patients, and grade 4 in 3 patients (Fig. 1). The MRC dyspnea scores at baseline were not different between the groups. MRC scores were significantly decreased in both groups after 6 months. There was no difference in MRC scores between the groups after 6 months.

Fourteen patients with restrictive lung diseases and 15 patients with COPD were receiving home oxygen therapy. PImax, PEmax, 6MWD, dyspnea, and emotional function subscales of the CRQ, and social functioning and role emotional subscales of the SF-36 increased significantly in the restrictive lung diseases group. In the COPD group, FVC (%predicted), PImax, PEmax, 6MWD, Borg score, dyspnea, fatigue, emotional function, and mastery subscales of the CRQ, and role emotional subscales of the SF-36 increased significantly (Tables 2 and 3). Repeated-measure ANOVA demonstrated no significant differences in improvement between the restrictive lung diseases group and the COPD group for any variable.

Discussion

In the present study, patients with restrictive lung diseases had increased PImax, PEmax, 6MWD, dyspnea and emotional function domains of CRQ, and social functioning and role emotional domains of SF-36, and decreased MRC scores, demonstrating that inspiratory and expiratory muscle forces, exercise tolerance, HRQOL and perception of dyspnea were improved following our home-based PR. There

Table 1. Baseline characteristics in restrictive lung disease and COPD patients.

	Restrictive lung disease	COPD	*P*
Age (years)	67 ± 14	70 ± 6	ns
Height (cm)	157 ± 15	160 ± 7	ns
Weight (kg)	50.6 ± 14.2	49.4 ± 10.0	ns
BMI (kg/m^2)	20.5 ± 4.2	19.3 ± 3.7	ns
FEV_1 (ml)	1.1 ± 0.5	1.1 ± 0.5	ns
FEV_1/FVC (%)	79.1 ± 10.4	43.0 ± 10.9	< 0.0001
FVC (% predicted)	53.4 ± 20.2	85.1 ± 22.0	< 0.0001
PImax (cmH_2O)	43.8 ± 22.8	45.8 ± 26.3	ns
PEmax (cmH2O)	60.6 ± 31.7	75.3 ± 30.1	ns
PaO_2 (mmHg)	73.9 ± 13.2	68.2 ± 14.8	ns
$PaCO_2$ (mmHg)	55.5 ± 8.7	43.0 ± 10.9	< 0.0001
6MWD (m)	308 ± 177	355 ± 131	ns
Borg score	4.5 ± 1.8	4.2 ± 1.5	ns

Data are expressed as means ± S.D. ns, not significant; BMI, body mass index; FEV_1, forced effort volume in one second; FVC, forced vital capacity; PImax, maximal inspiratory muscle forces; PEmax, maximal expiratory muscle forces; 6MWD, 6-minute walking distance.

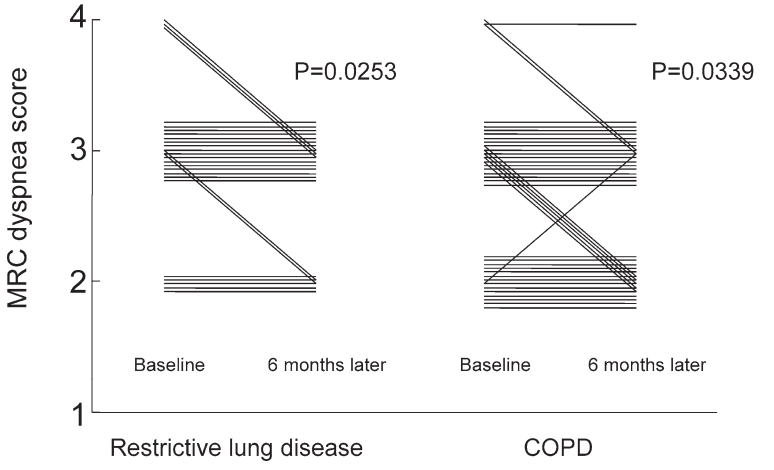

Fig. 1. Changes in Medical Research Council (MRC) dyspnea scores. Each line represents a patient with restrictive lung disease or COPD. There was no significant difference in MRC scores between the groups at baseline and after 6 months. Note that MRC scores were significantly decreased in both groups after 6 months.

were no differences in age, BMI, FEV_1, PImax, PEmax, PaO_2, 6MWD, Borg scale, and MRC scores at baseline between restrictive lung diseases patients and COPD patients. Although $PaCO_2$ was higher in restrictive lung diseases patients than in COPD patients, we assumed that the severity of disease for both groups was similar. Accordingly, our results also demonstrated that PR for the restrictive lung diseases group had the same effect as for the COPD group if the severity of the disease was similar.

Home-based PR is advantageous in that it requires minimal equipment, is less costly, and gives patients more time to spend with their families (Hernández et al. 2000), although it tends to be of a lower training intensity as a result of the limited supervision (Gosselink et al. 1997). Low-intensity exercise or muscle training in COPD patients resulted in modest improvements during a submaximal exercise test, while high-intensity training resulted in improvements during both maximal and submaximal exercise tests (Gosselink et al. 1997). However, activities of daily life require submaximal effort more so than they require maximal effort. Thus, from a practical viewpoint, low-intensity PR, which is easier to perform, may lead to improved long-term adherence (Normandin et al. 2002). In COPD patients, 12 weeks of home-based PR was shown to improve exercise capacity and the Borg scale, which were then maintained longer than when using hospital-based outpatient PR (Strijbos et al. 1996), while low intensity peripheral muscle training at home and once a week under the supervision of a physiotherapist in the hospital was reported to improve exercise tolerance and breathlessness (Clark et

Table 2. Changes in the parameters after 6 months in patients with restrictive lung disease and COPD.

	Restrictive lung disease		COPD	
Variables	6 months later	P^*	6 months later	P^*
Weight (kg)	49.4 ± 14.9	ns	51.3 ± 10.9	ns
BMI (kg/m^2)	20.2 ± 4.0	ns	20.0 ± 4.5	ns
FEV_1 (ml)	1.1 ± 0.5	ns	1.2 ± 0.5	ns
FEV_1/FVC (%)	72.2 ± 15.3	ns	42.9 ± 11.1	ns
FVC (% predicted)	55.0 ± 22.8	ns	93.0 ± 23.4	0.0016
PImax (cmH_2O)	57.2 ± 35.0	0.0215	57.4 ± 35.9	0.0069
PEmax (cmH2O)	84.3 ± 38.1	0.0003	92.9 ± 30.4	< 0.0001
6MWD (m)	354 ± 131	0.0362	391 ± 124	0.0167
Borg score	4.0 ± 1.2	ns	3.3 ± 1.6	0.0292

Data are expressed as means ± S.D. ns, not significant; BMI, body mass index; FEV_1, forced effort volume in one second; FVC, forced vital capacity; PImax, maximal inspiratory muscle forces; PEmax, maximal expiratory muscle forces; 6MWD, 6-minute walking distance. *compared to baseline.

Table 3. Changes in CRQ and SF-36.

		Restrictive lung disease			COPD		
Scale	Subscale	Baseline	6 months later	P	Baseline	6 months later	P
CRQ	dyspnea	20.0 ± 5.9	25.4 ± 5.1	0.0012	21.9 ± 6.2	26.1 ± 6.3	0.0005
	fatigue	18.7 ± 5.7	20.3 ± 4.8	ns	19.1 ± 5.6	22.2 ± 4.8	0.0072
	emotional function	36.5 ± 8.5	40.6 ± 6.4	0.0201	34.5 ± 8.6	40.0 ± 9.0	0.0013
	mastery	20.9 ± 6.1	22.3 ± 5.0	ns	19.4 ± 5.4	23.8 ± 5.0	0.0005
SF-36	Physical functioning	54.2 ± 27.3	60.0 ± 27.5	ns	56.8 ± 24.0	63.6 ± 23.5	ns
	Role physical	22.9 ± 39.1	51.7 ± 42.7	ns	52.5 ± 36.2	56.0 ± 44.1	ns
	Bodily pain	65.0 ± 30.3	68.0 ± 24.8	ns	71.5 ± 25.8	77.6 ± 24.2	ns
	General health	30.0 ± 16.1	39.7 ± 18.2	ns	44.2 ± 19.0	52.4 ± 19.6	ns
	Vitality	52.5 ± 21.8	55.0 ± 25.7	ns	58.2 ± 26.2	68.4 ± 20.9	ns
	Social functioning	69.8 ± 29.9	81.7 ± 23.1	0.0422	73.1 ± 28.5	77.2 ± 25.7	ns
	Role emotional	36.1 ± 43.7	82.2 ± 35.3	0.0384	53.3 ± 46.4	72.4 ± 45.5	0.0461
	Mental health	68.1 ± 23.5	71.2 ± 23.5	ns	64.2 ± 20.7	75.2 ± 21.6	ns

Data are expressed as means ± S.D. ns, not significant; CRQ, Chronic Respiratory Disease Questionnaire; SF-36, Short-Form 36

al. 1996). The results from the present study demonstrated that home-based PR is also effective in patients with restrictive lung disease.

Improvement in HRQOL is very important for chronic respiratory disease patients. Ferreira et al. (2006) reported that all four domains of CRQ significantly increased in non-COPD patients following PR. By contrast, in the present study dyspnea and emotional function domains increased significantly in restrictive lung disease patients following PR. These differences may be because the non-COPD patients in the Ferreira et al. (2006) study included obstructive lung disease other than COPD; approximately one-quarter of patients were asthmatic. A disease specific questionnaire such as CRQ is more sensitive to change than a non-disease specific questionnaire, although the validity and reliability of CRQ remains to be proven for patients with non-COPD. We simultaneously evaluated SF-36 as a non-disease specific evaluation, and observed a significant improvement in social functioning and role emotional in restrictive lung diseases patients, while only role emotional improved significantly in COPD patients. These data provide support for the use of PR in restrictive lung disease patients to improve HRQOL.

There were several limitations of the present study. First, we included only patients who completed PR for 6 months. Patients who dropped out within 6 months might not have had positive effects. Patients with MRC 5 were not included, probably as we excluded patients who could not visit our hospital every 2 weeks. It is unclear whether MRC 5 patients with restrictive lung disease may benefit from PR. The present study was not randomized and the sample sizes were small. In the future, a randomized con-

trolled study is required to clarify the benefits of PR for restrictive lung disease patients.

In conclusion, our home-based PR significantly improves inspiratory and expiratory muscle forces, exercise tolerance, HRQOL and perception of dyspnea in patients with restrictive lung disease to the same extent as in COPD patients.

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