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MEDICAL SCIENCES / DAHİLİ TIP BİLİMLERİ

Efficacy of Comprehensive Pulmonary Rehabilitation in Patients with Kyphoscoliosis; in Which Patients, the Gainings are Prominent?

Kifoskolyozisli Hastalarda Kapsamlı Pulmoner Rehabilitasyonun Etkinliği; Hangi Hastalarda Kazanımlar Belirgindir?

Seher Satar, Pınar Ergün

Ankara Atatürk Sanatorium Training and Research Hospital, Ankara, Turkey

Abstract

Objectives: Kyphoscoliosis (KS) is a common spinal deformity. The most common symptoms are dyspnea, reduced exercise capacity, and impaired quality of life. The aim of our study was to investigate the effectiveness of pulmonary rehabilitation (PR) in patients with KS and in which patients the gainings are prominent.

Materials and Methods: Forty-six patients who were diagnosed as KS and completed a multidisciplinary comprehensive supervised outpatient PR program were evaluated. Pulmonary functions, body composition, exercise capacity, respiratory and peripheral muscle strength, quality of life, psychological states were assessed before and after the PR program. The data of the patients were reviewed retrospectively.

Results: After the PR program, statistically significant differences were observed in Medical Research Council (MRC) levels, Borg, incremental shuttle walking test, endurance shuttle walking test, six minutes walk test, maximal inspiratory pressure, maximal expiratory pressure, hand grip test, deltoid and quadriceps 1-repetition maximum tests, St. George Respiratory Questionnaire (SGRQ), chronic respiratory diseases questionnaire, hospital anxiety and depression scores (HAD-anxiety/depression). There was a statistically significant negative correlation between the initial FEV_1 and the gain in anxiety. We also found a statistically significant negative correlation between the initial MRC, SGRQ, anxiety values and the gainings in depression data.

Conclusion: After multidisciplinary comprehensive PR program, there was an improvement in perception of dyspnea, muscle strengths, exercise capacities, quality of life, anxiety and depression scores. Our results indicate that the most prominent gainings were in the early stages of the disease.

Key Words: Kyphoscoliosis, Pulmonary Rehabilitation, Efficacy

Öz

Amaç: Kifoskolyoz (KS) sık görülen bir spinal deformitedir. En sık görülen semptomlar nefes darlığı, azalmış egzersiz kapasitesi ve bozulmuş yaşam kalitesidir. Çalışmamızın amacı, KS'li hastalarda pulmoner rehabilitasyonun (PR) etkinliğini ve kazanımların hangi hastalarda belirgin olduğunu araştırmaktır.

Gereç ve Yöntem: KS tanısı alan ve multidisipliner, kapsamlı, direkt gözetimli, ayaktan PR programını tamamlayan 46 hasta değerlendirildi. PR programı öncesi ve sonrasında solunum fonksiyonları, vücut kompozisyonu, egzersiz kapasitesi, solunum ve periferik kas kuvveti, yaşam kalitesi, psikolojik durumları değerlendirildi. Hastaların verileri geriye dönük olarak incelendi.

Bulgular: PR programından sonra; Medical Research Council (MRC) seviyeleri, Borg, artan hızda mekik yürüme testi, dayanıklılık mekik yürüme testi, altı dakika yürüme testi, maksimum inspiratuvar basınç, maksimum ekspiratuvar basınç, el kavrama testi, deltoid ve kuadriseps 1-tekrar maksimum testleri, St. George Solunum Anketi (SGRQ), kronik solunum hastalıkları anketi, hastane anksiyete ve depresyon (HAD-anksiyete/depresyon)

Address for Correspondence/Yazışma Adresi: Seher Satar
Ankara Atatürk Sanatorium Training and Research Hospital, Ankara, Turkey
Phone: +90 505 825 23 03 **E-mail:** sehersatar@yahoo.com **ORCID ID:** orcid.org/0000-0003-2896-6723
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skorlarında istatistiksel olarak anlamlı farklılıklar gözlendi. Başlangıç FEV_1 ile anksiyetedeki kazanım arasında istatistiksel olarak anlamlı bir negatif korelasyon vardı. Ayrıca başlangıçtaki MRC, SGRQ ve anksiyete değerleri ile depresyon düzeyi kazanımları arasında istatistiksel olarak anlamlı bir negatif korelasyon bulundu.

Sonuç: Multidisipliner kapsamlı PR programından sonra dispne algısı, kas kuvvetleri, egzersiz kapasiteleri, yaşam kalitesi, anksiyete ve depresyon puanlarında iyileşme oldu. Sonuçlarımız, en belirgin kazanımların hastalığın erken evrelerinde olduğunu göstermektedir.

Anahtar Kelimeler: Kifoskolyozis, Pulmoner Rehabilitasyon, Etkinlik

Introduction

One of the causes of the restrictive lung diseases is kyphoscoliosis (KS) which affects about 1% of the population but only 10% of the cases present significant clinical symptoms (1).

As a result of the combined effects of kyphosis, scoliosis and spinal rotation, the compliance of the chest wall decreases and the elastic recoil pressures of the chest wall and respiratory system increase. Patients are at a greater risk of muscle fatigue with aging, thereby reducing their chest wall compliance. So the breathing patterns of these patients change leading to hypoventilation, hipoxemia, hipercapnia and eventually leading to chronic respiratory failure (CRF) (2). After all, these patients develop shortness of breath during exercise, which is the most disturbing symptom, causing a decrease in exercise tolerance, limitation of daily living activities and impairment in health-related quality of life (HRQL) (2,3).

For all these reasons, KS is an important chronic clinical problem; but there is still no definitive guideline for these patients' management, particularly with respect to respiratory pathology caused by the disease. In the study of McMaster et al. (4), 41 patients who were operated were studied retrospectively. Twenty two patients (54%) had an impairment of respiratory function and finally they found that an increasing severity of kyphosis was associated with a significant increase in respiratory impairment ($p<0.005$). A major impairment of lung function is often observed when the kyphotic parts of the deformity predominate, probably caused by forward angulation of the spine and compression of the ribs on bothsides, thus impairing the movements of the diaphragm (5). Although many studies have mentioned that adding non-invasive mechanical ventilation (NIMV) and long-term oxygen treatment (LTOT) to treatment contributes positively to blood gas values, exercise capacity, HRQL and survival. Adding PR to the treatments of patients with or without respiratory failure provides the same benefits (5-10). KS with persistent symptoms like dyspnea, fatigue and/or functional status limitations despite optimal therapy, as stated in the American Thoracic Society-European Respiratory Society (ATS/ERS) statement, is one of the restrictive diseases that must be referred to PR (10,11). Although there is a small amount of data about pulmonary rehabilitation efficacy in patients with chest wall deformities, individualization of targets by creating patient-specific programs will increase PR effectiveness. In our study, our primary end point was to determine the effect of PR on dyspnea perception, exercise capacity, and quality of life in patients with symptomatic KS while our second end point was to determine the predictors of success according to the initial parameters.

Materials and Methods

Study Population

Between December 2005 and March 2020, sixty five patients with a diagnosis of KS were referred to our multidisciplinary pulmonary rehabilitation centre. Nineteen of the 65 patients were unable to complete the program. So 46 patients who completed the program were included in the study. The University of Health Sciences Turkey, Ankara Atatürk Chest Diseases and Thoracic Surgery Training and Research Hospital Ethics Committee, has approved the study (dated: 15.10.2020 and numbered 697-5) and the informed consent was obtained from the patients.

Exclusion Criterias

Patients having missing datas, and patients who dropped from the program (because of active work life, transportation or compatibility problems).

Outcome Parameters

Pulmonary functions, dyspnea perception, exercise capacity, respiratory and peripheric muscle srength, quality of life, body composition, and psychological status before and after the eight week outpatient PR program were recorded. Spirometry was performed to determine forced vital capacity (FVC), forced expiratory volume in one second (FEV_1) and FEV_1/FVC using a spirometer (AS-507, Minato Medical Science, Tokyo, Japan), in accordance with the ATS-ERS guidelines (12). Respiratory muscle strength was evaluated by measuring the maximal inspiratory pressure (MIP) and maximal expiratory pressure (MEP) using a Micro-RPM respiratory pressure meter (CareFusion, Hoechberg, Germany). MIP and MEP were measured with the subject in a sitting position by the same physiotherapist, in accordance with the recommendations of the ATS-ERS (13). MIP was measured starting from residual volume and MEP was measured starting from total lung capacity. Tests were repeated a minimum three times, and the best value was recorded. A hand grip test was

performed, measured using a hand dynamometer, to assess the peripheral muscle strength of the patients. Also 1-repetition maximum (1-RM) test is used to evaluate the peripheral muscle strength. Exercise capacity was evaluated using the incremental shuttle walking test (ISWT) and endurance shuttle walking test (ESWT). The tests were performed according to field walking tests guidelines (14,15). A value of minimal clinically important difference has been identified for the ISWT as 47.5 m (~5 shuttles) (14). In patients with severe dyspnea; the six-minute walk test (6MWT) was used. HRQL was assessed using the St. George's respiratory questionnaire (SGRQ) (16) and chronic respiratory diseases questionnaire (CRDQ) (17). Dyspnea was assessed using the MRC scale (18). Bioelectrical impedance was used to assess body composition using a TANITA (TBF-300A Total Body Composition Analyzer, Tokyo, Japan). Body mass index (BMI) and fat-free mass index (FFMI) were calculated using the formula of weight (body mass for BMI, fat-free mass for FFMI) in kilograms divided by the square of the height in meters. Hospital anxiety and depression (HAD) scores were used to assess psychological status (19).

Pulmonary Rehabilitation Program

Patients were evaluated by the chest physicians, physiotherapist, psychologist, dietician and a nurse who were part of the multidisciplinary team after a detailed medical history information and physical examination were obtained by the responsible chest physicians. After all the datas of patients were discussed in our multidisciplinary council, an individualized PR program was created according to the needs of each patient. Then patients underwent an 8-week hospital based outpatient comprehensive PR program and attended the rehabilitation unit on two half-days per week. The PR program consisted of exercise training, education, nutritional and psychosocial counseling. Structured educational sessions were provided monthly by a chest physician, two physiotherapists, a dietician, a nurse, and a psychologist and consists of disease education, self-management strategies, importance of exercise training, breathing retraining, airway clearance techniques, energy conservation techniques, medication advice, dietary advice, and psychosocial issues. Individual educational sessions were performed each follow-ups according to patients' needs.

The exercise training program consisted of eight weeks of endurance and resistance training. Inspiratory and expiratory muscle training were added when necessary. Programs were tailored to the individual needs, ability to tolerate exercise and disease severity.

The endurance training included 30 minutes (min) of endurance exercise (15 min on a treadmill and 15 min on a stationary bicycle) at 85% of each patient's VO_2 peak calculated from the ISWT. A 15-minute warm-up and cool-down period were also included. Quadriceps resistance training entailed leg extensions using free weights two days per week for eight weeks according to 1-RM, starting at 45% for two sets (10 repetitions per set) followed by 70% for three sets. Resistance training of the shoulder girdle and elbow muscle consisted of one set at 10 repetitions per set started at half a kilogram before being increased to 1-1.5 kg. The training was based on the recommendations of guidelines (10). Respiratory failure in KS has been most associated with respiratory muscle fatigue, which is why respiratory muscle strengthening and endurance-enhancing exercises are included in the program, especially in patients with inspiratory muscle weakness. IMT was applied to these patients by a inspiratory load of 30-50% maximum inspiratory pressure for 5-10 minutes, twice a day. In expiratory muscle traning in patients with low MEP values, the "rule of fives" is applied which consists of five exhalations and five blocks, equivalent to 75% of the MEP capacity of the patient per session, and five sessions per week for 5 weeks is recommended. During the PR sessions heart rate, blood pressure values and oxygen saturations of the patients were monitored by physiotherapists. Oxygen support was provided to keep the oxygen level above 90% if necessary.

Statistical Analysis

In our study, categorical independent data were analyzed using the chi-square test. Normal distribution of numeric variables was evaluated with Kolmogrov-Smirnov test, Skewness and Kurtosis. Normally-distributed numeric variables were expressed as mean and standard variation while non-normally distributed variables were expressed as median. The correlation between two numeric variables with normal distribution and a linear correlation was analyzed using Pearson's test. The correlation between variables that did not show normal distribution was evaluated with Spearman's test.

All analyses were performed using SPSS 22.0 statistical software (SPSS Inc, Chicago, IL, USA). $P<0.05$ was considered statistically significant.

Results

Sixty-five patients diagnosed with KS were evaluated for PR program but 19 (29%) of them dropped from the program for various reasons. Twenty-five (54%) of the forty-six patients with KS were male and twenty-one (46%) were female. The mean age of all patients was 52±13 years. Thirty-two (70%) of the patients never smoked, twelve (26%) were ex-smokers, while two (4%) were active smokers. While twenty-four (52%) of the patients using long term oxygen therapy, twenty two (48%) patients did not need oxygen support. All demographic characteristics and initial assessment parameters of the patients were given in Table 1.

A statistically significant difference was found between pre-PR and post-PR values in all MRC (p=0.000), Borg at rest (p=0.026), 6MWT (p=0.007), ISWT (p=0.000), ESWT (p=0.012), ESWT effectiveness (p=0.001), hand grip test (right/left) (p=0.000/p=0.000), deltoid and quadriceps 1RM test (right / left) (p=0.000), SGRQ (p=0.000), CRDQ (p=0.000), and HAD scores (p=0.000). The mean values of MIP measured before PR were 48.5±25 cmH_2O while it was 58±24 cmH_2O at the end of PR, a statistically significant difference was detected (p=0.000). Also, MEP values were measured as 86±43 cmH_2O and 100.5±41.5 cmH_2O before and after PR and there was a significant difference (p=0.000). To evaluate the peripheral muscle strength, hand grip test and 1RM were performed. Mean values in both right and left hand grip tests increased, with a statistically significant difference, compared to baseline after PR (p=0.001, p=0.001 respectively). Also 1RM results were statistically significant for both deltoid and quadriceps muscles. Furthermore no statistically significant difference was observed in FVC (p=0.230), FEV_1 (p=0.094), BMI and FFMI (p=0.889 and p=0.0394, respectively) (Table 2).

Table 1: Demographic features and initial assessment parameters

	Mean ± SD/Median (min:max)/number (%)
Age (years)	52±13/53 (26:76)
Gender (m/f) n (%)	25 (54%)/21 (46%)
Smoking(never/current/ex-smoker) n (%)	32 (70%)/2 (4%) / 12 (26%)
Smoking (p/year)	7.7±15.2/0 (0:70)
LTOT n (%)	24 (52%)
NIMV n (%)	19 (37%)
FEV_1 predicted %	33±12/32 (13:63)
FVC predicted %	35±16/32 (15:95)
MRC score	3±1/3 (1:5)
Borg at rest	0.3±0.5/0 (0:2)
Borg after exercise	3.1±1.3/3 (0:5)
ISWT (meters)	219±112/195 (40:430)
6MWT (meters)	244±108/290 (60:390)
ESWT (min.)	7±6/5 (0:20)
MIP (cmH_2O)	48±24/42 (17:133)
MEP (cmH_2O)	85±44/83 (21:187)
Hand grip test - Right	25±11/26 (8:54)
Hand grip test - Left	22±10/20 (6:42)
Deltoid 1-repetition maximum (kg)	4.3±1.7/4 (1:8)
Quadriceps 1-repetition maximum (kg)	8.2±3.2/8 (2:15)
BMI (kg/m^2)	26±5/26 (18:40)
FFMI (kg/m^2)	19±2/19 (16:24)
SGRQ total score	62.5±16/62 (29:100)
CRDQ	71±14/71 (26:106)
HAD - Anxiety scores	10±2/10 (7:14)
HAD - Depression scores	9±2/9 (6:14)

6MWT: Six minutes walk test, BMI: Body mass index, CRDQ: Chronic respiratory diseases questionnaire, ESWT: Endurance shuttle walking test, FEV_1: Forced expiratory volume in one second, FFMI: Fat-free mass index, FVC: Forced vital capacity, HAD: Hospital anxiety and depression, ISWT: Incremental shuttle walking test, kg: Kilogram, LTOT: Long term oxygen treatment, m/f: Male/female, max: Maximum, MEP: Maximal expiratory pressure, min:max: Minimum:maximum, min: Minute, MIP: Maximal inspiratory pressure, MRC: Medical research council, n: Number, NIMV: Non-invasive mechanical ventilation, p/year: Packet/year, SGRQ: St. George's respiratory questionnaire

A significant negative correlation was found between baseline FEV_1 values and gainings in anxiety scores (p=0.005, r=-0.432). Significant negative correlations were found between baseline MRC values and gainings in MRC and depression scores (p=0.09, r=-0.390). Similar to MRC, significant negative correlations were found between baseline SGRQ values and gainings in SGRQ and depression scores. Statistically significant negative correlation was observed in the initial and gainings of the MIP values (p=0.045, r=-0.341). And finally, a statistically significant negative correlation was found between the initial anxiety values and the gainings in anxiety and depression scores (p=0.000, r=-0.594; p=0.001, r=-0.470; relatively). The gainings and the comparisons with the initial assessment parameters were all shown in Table 3.

Discussion

In this study, we aimed to investigate the contribution of comprehensive multidisciplinary outpatient supervised pulmonary rehabilitation program in patients with KS. We found that there were statistically significant improvements in our patients' dyspnea perception, exercise capacity, respiratory and peripheral muscle strength, quality of life and psychosocial status. Also, pulmonary rehabilitation scores are more prominent in less symptomatic patients.

KS is one of the most serious chest wall abnormalities that affects breathing and exercise tolerance (20-22), and dyspnea is the common symptom. During exercise, decreases in inspiratory capacity, lung ventilation, and arterial oxygen saturation are common (23). Our patients' FEV_1 and FVC values both decreased as a result of this significant restriction, also their dyspnea levels, as determined by MRC, ranged from 2 to 4, and the Borg values indicated severe dyspnea after exercise. The higher reduction was detected in MIP values at the initial assessment, despite the fact that it was unrelated to the dyspnea scores.

The importance of PR in the treatment of chronic obstructive pulmonary disease (COPD) has been mentioned in numerous publications and guidelines to date, but its impact in non-COPD patients has yet to be recognized. In particular, studies on the effect of PR in patients with KS are very limited. In a

recent study conducted with 214 non-COPD patients, hospital-based PR was applied and statistically significant improvement was observed in 6MWT and SGRQ values (24). Cejudo et al. (25) demonstrated that patients with CRF due to KS improved significantly more in peripheral muscle strength, dyspnea, and quality of life when they participated in an exercise program. The authors claim that in KS, impairment in inspiratory muscle performance is linked to the development of respiratory failure. In another research, physicians measured transdiaphragmatic pressure during normal breathing and MIP in 9 KS patients to determine inspiratory muscle performance (26). MIP and transdiaphragmatic pressures were found to be significantly lower in the study participants than in the general population. That's why inspiratory muscle training (IMT) training should be implented in PR program when necessary. Our patients' mean MIP values were likewise low, thus those with low MIP values were given IMT, which consisted of a 5-10 minute inspiratory load of 30-50% maximum inspiratory pressure twice a day. Finally, we discovered a statistically significant improvement in their MIP values after respiratory muscle training. Twenty-four of our patients used LTOT, whereas 19 used NIMV. Our patients used LTOT for 16-18 hours per day on average, and NIMV for 4-6 hours per day, and all night during sleep. In the initial PR evaluation of all patients using LTOT and NIMV, arterial blood gas samples were obtained and examined, and all were determined to be compensated.

The underlying cause of many problems in KS, such as reduced exercise capacity and poor quality of life, is peripheral muscle weakness, which should be explored. Similar to Cejudo et al. (25), quadriceps weakness was hypothesized to be a feature of restrictive lung disease caused by scoliosis in another study, with the main conclusion being that quadriceps muscle weakness is linked to decreased exercise performance in these individuals as measured by the 6MWT (27). The association between hand grip strength and total muscle strength was explored in a different research with 384 healthy people, and a statistically significant relationship was discovered between hand grip strength and total muscle strength (28). After PR, we saw a statistically significant increase in peripheral muscle strength as determined by the all-hand grip test and 1RM results of both the deltoid and quadriceps muscles, which matched the findings of the previous studies.

Many variables, including ventilatory failure, contribute to the reduction of exercise capacity, in addition to peripheral muscle weakness. The findings of the 6MWT, shuttle walk, and maximum cycle test in patients with CS are limited (2,6,29). In a research by Fuschillo et al. (22), 18 individuals were given a PR program that lasted 4-6 weeks and saw a significant

Table 2: Outcome measures before and after the PR program

Parameters	Number	Before PR	After PR	P
FEV_1 predicted %	39	33.5±12.0	39.2±26.3	0.094
FVC predicted %	39	34.2±15.8	37.1±17.2	0.230
MRC score	45	3.1±0.9	2.6±0.8	**0.000**
Borg at rest	42	0.3±0.5	0.1±0.4	**0.026**
Borg after exercise	40	3.4±1.1	3.1±1.3	0.142
ISWT (meters)	30	224.7±113.5	287.7±119.5	**0.000**
6MWT (meters)	11	244.3±107.8	284.5±101.9	**0.007**
ESWT (min.)	30	6.8±5.9	13.9±7.7	**0.001**
MIP (cmH_2O)	35	48.5±24.9	57.8±23.9	**0.000**
MEP (cmH_2O)	35	86.0±43.1	100.5±41.5	**0.000**
Handgrip test - Right	37	24.9 ±10.8	27.5±10.5	**0.000**
Handgrip test - Left	37	22.4±25.3	22.4±10.1	**0.000**
Deltoid 1RM (kg)	35	4.3±1.7	4.8±1.7	**0.000**
Quadriceps 1RM (kg)	34	8.3±3.2	9.4±2.9	**0.000**
BMI (kg/m^2)	45	25.9±5.4	25.9±5.1	0.889
FFMI (kg/m^2)	44	19.1±2.0	19.0±1.9	0.394
SGRQ Score	45	60.9±18.5	35.4±13.5	**0.000**
CRDQ Score	45	14.7±4.3	19.9±5.1	**0.000**
HAD - Anxiety	44	9.9±1.6	7.0±1.4	**0.000**
HAD - Depression	44	9.4±1.9	7.3±1.3	**0.000**

1RM: 1-repetition maximum, 6MWT: Six minutes walk test, BMI: Body mass index, CRDQ: Chronic respiratory diseases questionnaire, ESWT: Endurance shuttle walking test, FEV_1: Forced expiratory volume in one second, FFMI: Fat-free mass index, FVC: Forced vital capacity, HAD: Hospital anxiety and depression, ISWT: Incremental shuttle walking test, kg: Kilogram, MEP: Maximal expiratory pressure, min: Minimum, min: Minute, MIP: Maximal inspiratory pressure, MRC: Medical research council, SGRQ: St. George's respiratory questionnaire

improvement in 6MWT distance (6MWD). Another study found statistically significant improvements in the endurance time and shuttle walking test in 34 KS patients with respiratory failure who were randomized into two groups with and without PR administered (25). In our study, patients with KS were underwent a comprehensive multidisciplinary 8-week PR program, and we observed statistically significant improvements in all of the ISWT, ESWT and 6MWD results.

Respiratory failure, dyspnea, and lower exercise capacity are the determinants of HRQL in chronic respiratory diseases. The CRDQ was frequently used to assess HRQL in individuals with respiratory failure and KS. In a study from Ireland, 46 patients

Table 3: The gainings and the comparison with the initial assessment parameters

		ΔMRC	ΔSGRQ	ΔISWT	ΔESWT	ΔMIP	ΔAnxiety	ΔDepression	ΔHand grip test	ΔDeltoid 1RM	ΔQuadriceps 1RM
Age	p	0.321	0.509	0.831	0.500	0.768	0.334	0.131	0.583	0.448	0.514
	r	-0.149	-0.100	-0.039	0.128	-0.051	0.149	0.231	0.093	0.131	-0.112
Gender	p	0.483	0.116	0.062	0.712	0.186	0.326	0.739	0.512	0.941	0.147
	r	-0.106	-0.235	-0.333	0.070	0.225	-0.152	-0.052	0.111	0.013	0.247
Smoking (pyear)	p	0.769	0.443	0.133	0.596	0.259	0.816	0.906	0.517	0.139	0.267
	r	0.044	0.116	0.271	-0.101	-0.193	0.036	-0.018	-0.110	0.252	-0.190
FEV_1	p	0.343	0.806	0.603	0.637	0.154	**0.005**	0.095	0.276	0.668	0.373
	r	-0.150	-0.039	-0.099	-0.093	-0.250	-0.432	-0.268	0.189	0.076	-0.158
FVC	p	0.628	0.560	0.999	0.518	0.498	0.759	0.338	0.171	0.809	0.419
	r	-0.077	-0.092	0.000	-0.127	-0.120	-0.050	-0.155	0.237	0.043	-0.143
MRC	p	**0.023**	0.116	0.294	0.877	0.878	0.771	**0.009**	0.854	0.338	0.185
	r	-0.335	-0.235	-0.191	-0.030	0.027	-0.045	-0.390	0.031	0.164	0.226
SGRQ	p	0.236	**0.000**	0.134	0.684	0.965	0.382	**0.002**	0.595	0.454	0.233
	r	-0.180	-0.604	-0.275	0.079	0.008	-0.135	-0.462	-0.092	0.129	0.204
CRDQ	p	0.268	**0.014**	0.198	0.711	0.942	0.756	**0.001**	0.362	0.644	0.804
	r	0.167	0.360	0.234	-0.070	-0.013	0.048	0.497	0.154	-0.080	-0.043
ISWT	p	0.106	0.562	0.950	0.698	0.747	0.128	0.770	0.880	0.072	0.190
	r	0.291	0.106	-0.012	-0.074	-0.066	-0.284	0.056	0.031	-0.345	-0.255
ESWT	p	0.086	0.555	0.815	0.268	0.978	0.245	0.972	0.493	0.309	0.917
	r	0.308	0.108	0.044	-0.209	0.006	-0.219	0.007	-0.141	-0.199	-0.021
MIP	p	0.979	0.466	0.491	0.935	**0.045**	0.648	0.961	0.320	0.512	0.565
	r	0.004	0.120	0.139	-0.017	-0.341	-0.077	0.008	-0.176	0.120	-0.106
MEP	p	0.837	0.934	0.996	0.588	**0.008**	0.870	0.982	0.090	0.582	0.685
	r	0.035	0.014	-0.001	0.111	-0.444	0.028	0.004	-0.295	0.103	-0.076
Anxiety	p	0.329	0.311	0.423	0.956	0.758	0.000	**0.001**	**0.034**	0.911	0.341
	r	0.149	-0.154	0.149	-0.011	-0.054	-0.594	-0.470	-0.354	0.019	-0.163
Depression	p	0.546	0.111	0.853	0.583	0.784	0.034	**0.000**	0.713	0.878	0.557
	r	-0.092	-0.241	0.035	-0.106	0.048	-0.320	-0.745	-0.063	-0.027	-0.101
Deltoid 1RM	p	0.449	0.764	0.689	0.885	**0.002**	0.305	0.192	0.351	0.164	**0.015**
	r	0.130	0.052	0.079	0.029	-0.528	0.176	0.222	0.170	-0.241	-0.410
Quadriceps 1RM	p	0.309	0.276	0.150	0.715	**0.008**	0.468	0.927	0.789	0.885	**0.024**
	r	0.177	0.189	0.284	0.074	-0.477	-0.127	-0.016	0.049	-0.025	-0.382

1RM: 1-repetition maximum, CRDQ: Chronic respiratory diseases questionnaire, ESWT: Endurance shuttle walking test, FEV_1: Forced expiratory volume in one second, FVC: Forced vital capacity, ISWT: Incremental shuttle walking test, MEP: Maximal expiratory pressure, MIP: Maximal inspiratory pressure, MRC: Medical research council, p/year: Packet/year, SGRQ: St. George's respiratory questionnaire

with restrictive lung disease (35 interstitial lung diseases, 11 skeletal abnormalities) were subjected to an 8 week PR program and followed for up to 1 year (30). A significant improvement was observed in dyspnea, quality of life and HAD scores of patients who completed the PR program. Also a significant decrease in the duration of hospitalization within 1 year following rehabilitation was observed. We both used CDRQ and SGRQ as HRQL indicators, and it was observed that there was a significant improvement in all parameters including total score and subheadings after PR. HAD scale was used to evaluate the patients' psychosocial status and a statistically significant improvement was observed both in anxiety and depression scores.

The secondary goal of this study was to determine the predictors of success based on the first assessment parameters. Anxiety levels were found to be much lower in patients with impaired respiratory functions. Patients with decreased dyspnea perceptions improved their depression scores more. Also, the depression scores improved when quality of life measures indicated a better state. The increases in inspiratory muscle strength were substantially more significant in patients with upper and lower limb muscle weakness. These results suggest an early referral is important for KS patients to a pulmonary rehabilitaion program.

Study Limitations

The limitations of this study can be evaluated as the use of patient data belonging to only our PR center and therefore the low number of patients. Other limitations are that the study was not a randomized controlled trial.

Conclusion

This study shows that the comprehensive multidisciplinary hospital-based outpatient pulmonary rehabilitation program in patients diagnosed with KS improved dyspnea perception, respiratory and peripheral muscle strength, exercise capacity with endurance time, HRQL, anxiety and depression levels. Considering that KS may present symptoms starting from a young age, patients with this diagnosis should be directed to PR centers without waiting for deterioration.

Ethics

Ethics Committee Approval: The University of Health Sciences Turkey, Ankara Atatürk Chest Diseases and Thoracic Surgery Training and Research Hospital Ethics Committee, has approved the study (dated: 15.10.2020 and numbered 697-5).

Informed Consent: The informed consent was obtained from the patients.

Peer-reviewed: Externally peer-reviewed.

Authorship Contributions

Concept: S.S., P.E., Design: S.S., P.E., Data Collection or Processing: S.S., P.E., Analysis or Interpretation: S.S., P.E., Literature Search: S.S., P.E., Writing: S.S., P.E.

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