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CASE REPORT

Effect of pulmonary rehabilitation on adult cystic fibrosis patient: a case report

Gamze YALÇINKAYA[1], Aylin Özgen APAYDIN[2], Sevgi ÖZALEVLI[3]

Cystic fibrosis is an autosomal recessive and fatal disease that affect the respiratory functions and daily living activities of patients. Fifty percent of the cystic fibrosis population consists of adult patients aged 18 years and older. The most common problems are decreased pulmonary function and exercise capacity in adult cystic fibrosis. In addition, many different comorbidities can be seen such as diabetes, osteoporosis and pain due to systematic influence. Pulmonary rehabilitation is known to increase mucus clearance and exercise tolerance in adult cystic fibrosis patients, however the studies that investigating the effectiveness of pulmonary rehabilitation are limited in this population. Therefore, pulmonary rehabilitation program of an adult patient with cystic fibrosis was performed in our study. The patient was included in an 8-week supervised pulmonary rehabilitation program with two sessions per week. The patient's severity of dyspnea perception, pulmonary function test, cardiopulmonary exercise test, static stabilization of the trunk muscles, quadriceps muscle strength, grip strength and health-related quality of life were assessed before and after the pulmonary rehabilitation program. Breathing exercises, bronchial hygiene techniques, aerobic training and home program were applied to our patient within the scope of pulmonary rehabilitation program. The patient's severity of dyspnea perception was decreased while 6-minute walking distance, cardiopulmonary exercise test parameters and respiratory function test results increased at the end of the 8-week supervised pulmonary rehabilitation program. The patient's grip strength and static trunk endurance increased while the quadriceps muscle strength did not change. In addition, it was determined that the patient's quality of life increased according to the Saint George Respiratory Questionnaire.

Keywords: Cystic Fibrosis, Rehabilitation, Exercise.

Pulmoner rehabilitasyonun erişkin kistik fibrozisli hasta üzerindeki etkisi: bir olgu sunumu

Kistik fibrozis otozomal resesif geçişli, hastaların solunum fonksiyonları ve günlük yaşam aktivitelerini etkileyen ölümcül bir hastalıktır. Kistik fibrozisli popülasyonun %50'si 18 yaş ve üzeri erişkin hastalardan oluşmaktadır. Erişkin kistik fibroziste en yaygın görülen problemler solunum fonksiyonları ve egzersiz kapasitesinin azalmasıdır. Ek olarak, sistematik etkilenimden dolayı diyabet, osteoporoz ve ağrı gibi birçok farklı problem de görülebilmektedir. Erişkin kistik fibrozisli hastalarda pulmoner rehabilitasyonun mukus klirensini ve egzersiz toleransını artırdığı bilinmektedir ancak bu popülasyonda pulmoner rehabilitasyonun etkinliğini araştıran yayınlar sınırlı sayıdadır. Bu nedenle, çalışmamızda kistik fibrozisli erişkin bir hastanın pulmoner rehabilitasyon programı gerçekleştirildi. Hasta, haftada iki seans olacak şekilde 8 haftalık hastaya özgü planlanmış bir pulmoner rehabilitasyon programına dahil edildi. Programının öncesinde ve sonrasında hastanın dispne seviyesi, solunum fonksiyon testi, kardiyopulmoner egzersiz testi, gövde kaslarının statik stabilizasyonu, quadriceps kas kuvveti, kavrama kuvveti ve sağlıkla ilişkili yaşam kalitesi değerlendirildi. Pulmoner rehabilitasyon programı kapsamında hastamıza solunum egzersizleri, bronşiyal hijyen teknikleri, aerobik eğitim ve ev programı uygulandı. 8 haftalık pulmoner rehabilitasyon programı sonunda hastanın 6 dakika yürüme mesafesi, kardiyopulmoner egzersiz testi parametreleri ve solunum fonksiyon test sonuçları artarken, algılanan dispne şiddeti azaldı. Hastanın quadriceps kas kuvvetinde tedavi başlangıcına göre değişim görülmezken, kavrama kuvveti ve statik gövde endurans arttı. Ek olarak, Saint George Solunum Anketi'ne göre hastanın yaşam kalitesinin arttığı belirlendi.

Anahtar kelimeler: Kistik Fibrozis, Rehabilitasyon, Egzersiz.

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1: Dokuz Eylul University, Institute of Healh Sciences, Izmir, Turkey
2: Dokuz Eylul University, Faculty of Medicine, Department of Pulmonary Diseases, Izmir, Turkey
3: Dokuz Eylul University, School of Physical Therapy and Rehabilitation, Izmir, Turkey
Corresponding author: Gamze Yalçınkaya: gamzeyalcinkaya11@gmail.com
ORCID IDs (order of authors): 0000-0003-2527-8191; 0000-0002-5711-1372; 0000-0002-5528-1036
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Cystic fibrosis (CF) is the autosomal recessive, progressive, multi-organ and life-shortening disease and the mean life expectancy of patients increase over the years. CF affects more young and adult populations and its present in 90.000 people in worldwide. 50 % of the CF population consists of adult patients aged 18 years and older.[1] Pulmonary rehabilitation (PR) programs have multiple beneficial effects in CF. Evidence suggests that exercise therapy contributes improving lung capacity and quality of life in CF. It also decreases the severity of dyspnea perception and increases exercise tolerance.[2] However, the studies investigating the effectiveness of PR in adult CF population are still limited. Evidence indicates that treadmill and cycle ergometer aerobic training increase ventilation-perfusion rate and provides mucus clearance. Exercise therapy also provide control of type 1 diabetes in adult CF patients through improving insulin sensitivity.[3,4] Adult CF patients may also experience comorbidities such as osteoporosis, diabetes, urinary incontinence and musculoskeletal problems due to multiple organ involvement. Pain, decreased peripheral muscle strength and influence of postural control are the musculoskeletal problems among adult CF patients. Lima et al. reported that imbalance between head, trunk and lower extremities may lead decreased quadriceps muscle strength in adult CF patients.[5] Despite all these multiple influences, there was no standardized pulmonary rehabilitation in adult CF patients. Therefore, we investigated the effects of PR program in a case of adult cystic fibrosis.

CASE

A 29-year-old female patient (54 kg, 172 cm, 18.25 kg/m^2) was diagnosed with CF when she was one-year-old. Patient's profession is teaching biology and she has no habit of smoking, alcohol or exercise. There is no comorbid disease in patient's medical history. However, patient's mother has type 2 diabetes mellitus. Patient has referred to the pulmonary rehabilitation unit due to progressive dyspnea, sputum (10 milliliter/day) and cough symptoms and decreased exercise capacity. The patient had an exacerbation period within the last year. Besides, one dose of 2.5 mg dornase alpha and 3 doses of 3 mg tobramycin drugs inhaled daily by the patient.

Evaluations

All evaluations were conducted at the beginning and end of the pulmonary rehabilitation program after receipt of informed consent form.

Severity of dyspnea: The dyspnea severity of the patient in daily life and during exercise were evaluated according to Modified Medical Research Council (MMRC) scale and modified Borg scale, respectively.[6]

Muscle Strength: Left and right quadriceps muscle strength were evaluated by hand-held dynamometer (Lafayette, England®). Left and right hand grip strengths were measured by hand grip dynamometer (Jamar, England ®).[7,8]

Static Trunk Stabilization Tests: Sorenson test, endurance test of trunk flexors, left and right-side bridge tests were followed for evaluating of static trunk stabilization endurance. All test performed twice and average total duration recorded in seconds.[9]

Pulmonary Function Test (PFT): Pulmonary function tests was performed on electronic Spirometer (JaegerTM, MasterScreen PFT) and its accompanying software (Sentry Suite version 2.19). The tests were carried out with the described procedure in ATS/ERS guidelines.[10] Obtained parameters were forced expiratory volume in first second (FEV1), forced vital capacity (FVC), FEV1/FVC% and peak expiratory flow (PEF).

Exercise Capacity: Six-minute walking test (6MWT) and cardiopulmonary exercise test (CPET) were evaluated to determine patient's exercise capacity. CPET evaluation were performed with bicycle ergometer by Bruce protocol in laboratory according to ATS/ERS guideline. The maximal oxygen consumption (VO2max), which the patient could reach in one minute, was evaluated with Ergomedic 839 E model bicycle ergometer (Monark, Sweden®) in the exercise laboratory. During the test, the oxygen ratio from the collected respiratory air by the one-way mask was measured by the metabolic analyzer system (Fitmate Pro, Cosmed, Italy®) and VO_2 levels were determined. The highest VO2 value measured at the highest work intensity was considered as aerobic capacity (VO_{2max}).[11]

Quality of Life: Patient's quality of life was assessed with Saint George's Respiratory

Questionnaire (SGRQ). Symptom, activity, impact and total scores were calculated respectively.[12]

In the first evaluation, the patient had sputum (10 milliliter/ day) and cough complaints. The patient's severity of dyspnea perception was also at stage two according to the MMRC scale. She was able to walk 75.13% (555 m) of her lower distances limit (599.64 m) according to Enright's reference value formula "Lower Limit= ((6MWD= (2.11 × height cm) − (2.29 × weight kg) - (5.78 × age) + 667 m)) - 139".[13] The patient's spirometric data (FEV1: 1.75 L/sec, FVC: 2.63 L/sec, FEV1/FVC: 66.51%) showed an obstructive pattern prior to the pulmonary rehabilitation program. CPET parameters (Load: 116 watts, VO_{2max}: 1141 ml/min/kg, SpO_2: 95%, Heart Rate: 160 beat/ min) and pre-PR functional assessments that including static trunk stabilization tests, quadriceps and grip strengths, and health-related quality of life scores were shown in Table 1.

Pulmonary rehabilitation program

The patient was participated in a 60-minute/ session, eight-week supervised PR program with two sessions per week. Thoracic expansion exercises and bronchial hygiene techniques (postural drainage) (15 min), treadmill and bicycle ergometer, aerobic training (30 min) and stretching exercises and diaphragmatic breathing exercises (15 min) were applied in every session. Treadmill (speed of 5.0 km/h and 5% incline) were adjusted that patient could reach 50% of her maximum heart rate according to CPET, after five minute warming period with walking speed (5.0 km/h) for the aerobic training. The exercise workload was increased by 10% when the patient was able to exercise for 10 min without any intolerable dyspnea measured as modified Borg Scale< 5 in each session. The cooling period had completed in five minutes for treadmill ergometer of aerobic training. Cycle-ergometer (Monark, Switzerland®) aerobic training was performed with pre-fixed intensity according to modified Borg Scale (dyspnea rating < 5) in for 10 min, following the water break and the control of physiological responses (heart rate, oxygen saturation- SpO_2 and blood pressure). In the last part, stretching exercises were performed five repetition/20 secs for left and right upper trapezius, quadriceps, hamstrings and lumbar extensor muscle groups. The PR session was completed with 10 diaphragmatic exercise. We did not use any equipment for oxygen assistance in all PR sessions. Addition to supervised PR program, the patient was followed by a three-day home exercise program. Thera band exercises were planned for the strengthening of shoulder flexors and abductors and 4-way hip exercises within the scope of the home exercise program. All home exercise program was asked to be performed three sets of 10 repetitions. The home program was completed by repeating the stretching of the same muscle groups as in the supervised PR program and 10 diaphragmatic exercises. The patient's participation in the home program was 83.3% at the end of eight weeks according to exercise diary. The patient also fully participated in all sessions of supervised PR program in the unit.

The patient reported that the sputum (5 milliliter/day), severity of dyspnea perception (MMRC: 0) and cough symptoms were decreased compared to the onset of the PR program. The six-minute walk distance increased to 792 meters that the patient had to walk according to Enright's formula (738.64 m).[13] Spirometry data of the patient (FEV1: 1.97 L/sec; FVC: 2.63 L/sec; FEV1/FVC: 74.32%) and CPET parameters (Load: 122 watts, VO_{2max}: 1334 ml/min/kg, SpO_2: 95%, Heart Rate: 171 beat/ min) were found to be increased compared to the beginning of the PR program. Additionally, the patient's quadriceps muscle strength did not change, but the grip strength and static trunk endurance increased. Health-related quality of life parameters especially activity scores of the patient also improved after eight weeks of treatment (Table 1).

DISCUSSION

Pulmonary rehabilitation is a necessary part of the treatment to maintain the general well-being of patient in chronic respiratory diseases. The European Society of Cystic Fibrosis reported that physiotherapy of CF should include airway cleaning techniques and exercise training in the current guideline.[14] However, the number of randomized controlled trials performed in adult CF is relatively low. Rovedder et al. reported that upper extremity muscle strength increased in supervised

Table 1. Changes in functional parameters before and after the pulmonary rehabilitation program.

	Pre-Pulmonary rehabilitation	Post-Pulmonary rehabilitation
Dyspnea (Modified Medical Research Council score)	2	0
Modified Borg Scale	3	1
6 Minute Walking Test (6MWT) (m)	555	792
Pulmonary Function Test		
FEV_1 (L/sec),(%)	1.75, 52	1.97, 58
FVC (L/sec),(%)	2.63, 68	2.63, 68
FEV_1/FVC (%)	66.51, 80	74.32, 89
PEF (L/sec),(%)	4.96, 67	4.99, 68
MEF25-75 (L/sec),(%)	0.34 - 3.16, 16-50	0.52 - 4.17, 24-66
Cardiopulmonary Exercise Test (at maximum Watts)		
Load (Watt)	116	122
VO_{2max} (ml/min/kg)	1141	1334
SpO_2 (%)	95	95
HR (beat/min)	160	171
Static Trunk Stabilization Tests		
Sorenson Test (sec)	29.90	32.37
Endurance Test of Trunk Flexors (sec)	59.40	146.04
Left Side Bridge Test (sec)	57.30	81.00
Right Side Bridge Test (sec)	52.35	82.20
Muscle strength (kg)		
Right quadriceps muscle strength (kg)	18.60	19.03
Left quadriceps muscle strength (kg)	18.90	18.90
Right hand grip strength (kg)	25.80	27.30
Left hand grip strength (kg)	24.30	28.00
Saint George's Respiratory Questionnaire		
Symptom score	40.70	18.10
Activity score	35.79	5.25
Impact score	40.76	19.03
Total score	39.24	22.09

FEV_1: Forced Expiratory Volume in First Second. FVC: Forced Vital Capacity. PEF: Peak Expiratory Flow. MEF: Mid-expiratory Flow Rate.
SpO_2: Peripheral Oxygen Saturation. HR: Heart Rate.

exercise group more than the home based exercise group, but there was no difference between the groups in terms of six minutes walking test and quality of life.[15] In our case report, supervised PR program increased the grip strength and six-minute walking test results report similar with aforementioned study. More recently, Beaudoin et al. stated that combining aerobic and strengthening exercises in adult CF patients with glucose intolerance developed glycemic control after 12 week exercise program.[3] In another report, both aerobic training and strength training were increased forced expiratory volume in first second (FEV_1) that similar with our findings.[16] Besides, interval training had effect VO_{2max} more than standard exercise protocol.[17] Thus, PR could increase the physiological parameters and quality of life of the adult CF patients. The present study also showed that 8-week supervised PR program and home-based exercises developed severity of dyspnea perception, six-minute walking distance, pulmonary functions, cardiopulmonary

parameters, grip strengths and quality of life in 29 year old adult CF case. The patient's severity of dyspnea perception might have decreased with the result of breathing exercises and bronchial hygiene techniques. Decreasing of dyspnea perception could increase the oxygenation of the patient by improving respiratory functions, muscle strengths and endurance. Therefore, the patient had able to tolerate more workload by similar SpO_2% value, heart rate and less dyspnea perception in the cardiopulmonary exercise test compared with the beginning of the PR program. Regulation of the dyspnea perception might also facilitate the participation of the patient in daily activities.

Patients with adult CF could experience problems such as skeletal muscle weakness, exercise intolerance, posture, urinary incontinence, participation in work life and quality of life.[18] For instance, 68% of women over 11 years of age have been reported to be affected by urinary incontinence.[19] The consideration of core stabilization as a part of the PR program could provide protective benefits against urinary incontinence, especially in female CF patients cause of the abdominal muscles and the pelvic floor connection.[20] Strengthening and aerobic exercise training increased static trunk muscles endurance in our case. Therefore, exercise training may also be efficient against the incontinence comorbidity which seen in advanced stages of adult CF patients.

Limitations

The literature recommends exercise for cystic fibrosis patient at least three times a week with 70-80% of VO_{2max}.[21] However, we carried out supervised pulmonary rehabilitation program of our case twice a week in order to comply with the transportation conditions of patient. This is the limitation of our study.

Conclusion

In conclusion, supervised pulmonary rehabilitation is a useful modality in the management of respiratory symptoms. Cohort studies with the larger group and definition of standardized rehabilitation could obtain more clinical information about benefit of physiotherapy program.

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