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KLİNİK ÇALIŞMA / ORIGINAL ARTICLE

Pulmoner arteriyel hipertansiyonlu bireylerde hastalık şiddetinin üst ekstremite kas kuvveti, egzersiz kapasitesi ve günlük yaşam aktivitelerine etkisi

Effect of disease severity on upper extremity muscle strength, exercise capacity, and activities of daily living in individuals with pulmonary arterial hypertension

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ÖZET

Amaç: Pulmoner arteriyel hipertansiyon (PAH) hastalarında kas kuvveti, egzersiz kapasitesi, yaşam kalitesi ve günlük yaşam aktivitelerinin etkilendiği bilinmesine rağmen hastalık şiddetinin ilerlemesi ile birlikte bunların nasıl etkilendiği bilinmemektedir. Bu çalışmada, hastalık şiddetinin PAH'lı bireylerde kas kuvveti, egzersiz kapasitesi ve üst ekstremite günlük yaşam aktivitelerine etkisinin incelenmesi amaçlandı. ***Yöntemler:*** Çalışmaya fonksiyonel sınıf düzeyi New York Kalp Cemiyeti (NYHA) sınıf II (n=14) ve III (n=11) olan 25 hasta alındı, hastalık şiddeti NYHA'ya göre sınıflandırıldı. Üst ekstremite egzersiz kapasitesini değerlendirmek için Altı Dakika Pegbo- ard ve Ring Testi (6PBRT) kullanıldı. Üst ekstremite günlük yaşam aktiviteleri kısıtlamalarını değerlendirmesi Milliken günlük yaşam aktiviteleri skalası (MAS) ile yapıldı. Omuz fleksiyonu, dirsek ekstansiyonu, dirsek fleksiyonu kas kuvveti ve el kavrama kuvveti dinamometreyle değerlendirildi.

Bulgular: Gruplar arasında yaş, cinsiyet, beden kitle indeksi ve ortalama pulmoner arter basınç özellikleri açısından farklılık bulunmadı (p>0.05). 6PBRT, MAS ve dirsek fleksiyonu (sağ) ve kavrama kuvvetleri (sağ ve sol) NYHA III'te NYHA II'ye göre anlamlı olarak düşük bulundu (sırasıyla, p=0.004, p=0.002, p=0.043, p=0.002 ve p=0.003). Omuz fleksiyonu, dirsek fleksiyonu (sol) ve dirsek ekstansiyonunda gruplar arasında anlamlı fark bulunmadı (p>0.05).

Sonuç: Bu çalışmanın sonuçları PAH'lı hastalarda hastalık şiddeti arttıkça üst ekstremite egzersiz kapasitesi, dirsek fleksiyonu kas kuvveti (sağ) ve kavrama kuvvetlerinin azaldığını ve üst ekstremite günlük yaşam aktivitesi kısıtlılıklarının arttığını düşündürmektedir. PAH hastalarında rehabilitasyon programları planlanırken hastalık şiddeti göz önünde bulundurulmalıdır ve üst ekstremitelere yönelik değerlendirmelere ve tedavilere yer verilmelidir.

ABSTRACT

Objective: Pulmonary arterial hypertension (PAH) is a rare disease. Although muscle strength, exercise capacity, quality of life, and activities of daily living of patients with PAH are affected, it is not known how they are affected by disease severity. The purpose of the present study was to investigate the effects of disease severity on upper extremity muscle strength, exercise capacity, and performance of activities of daily living in patients with PAH.

Methods: Twenty-five patients with disease severity classified according to the New York Heart Association (NYHA) as functional class II (n=14) or class III (n=11) were included in the study. Upper-extremity exercise capacity and limitations in performing activities of daily living were assessed with the 6-minute Pegboard and Ring Test (6PBRT) and the Milliken Activities of Daily Living Scale (MAS), respectively. Shoulder flexion, elbow extension, elbow flexion muscle strength, and handgrip strength were measured with dynamometer.

Results: There were no significant differences in age, gender, body mass index, or mean pulmonary artery pressure between groups (p>0.05). The 6PBRT, MAS, and elbow flexion (right) and grip strength (right and left) results were significantly lower in NYHA III group than in NYHA II group (p=0.004, p=0.002, p=0.043, p=0.002 and p=0.003, respectively). There was no significant difference in shoulder flexion, elbow flexion (left), or elbow extension between groups (p>0.05).

Conclusion: Results suggest that upper extremity exercise capacity, elbow flexor muscle strength (right), and handgrip strength decrease and that limitations in activities of daily liv¬ing grow as disease severity increases in patients with PAH. When planning rehabilitation programs, disease severity should be considered and evaluations and treatments for the upper extremities should be included.

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Pulmonary arterial hypertension (PAH) is a rarely seen disease considered as a subgroup of hypertension.[1] PAH is clinically classified in four categories as idiopathic, heritable, drugs and toxins induced, and associated with other diseases (connective tissue disease, HIV infection, portal hypertension, congenital heart disease, and schistosomiasis).[2] PAH causes right ventricular insufficiency, and cardiopulmonary dysfunction.[3] In patients with PAH, dyspnea, fatigue, and chest pain are important symptoms.[34] These symptoms impair exercise capacity, physical functions, and quality of life.[3]

Loss of strength of peripheral muscles is one of the important causes of restricted exercise capacity.[5] Important role of peripheral muscles in the restriction of exercise has been demonstrated in patients with PAH.[6,7] It has been thought that poor oxygenation as a result of PAH, strength of peripheral muscles decreases, in parallel with the disease progression, exercise capacity is restricted in patients with PAH.[8] In patients with PAH, decrease in cross-sectional area of type 1 fibers, has been found to induce changes in muscle functions, and decrease in force–producing capacity when compared with healthy controls.[9] In patients with PAH, decreases in peak oxygen intake, anaerobic threshold, ventilatory effectiveness, and 6-Min Walk Distance (6MWD) were detected.[1011] The severity of these changes increases with progression in functional New York Heart Association (NYHA) classification which is very frequently used to classify disease severity. [12,13]

Upper extremities play important roles in performing of many activities of daily living such as eating, personal hygiene, and vocational tasks.[14] Functional impairment of upper extremities adversely compromise level of independence of the individuals, and effect their quality of life.[14] The results of studies in patients with chronic obstructive pulmonary disease (COPD) have indicated that patients are experiencing problems with activities of daily living, especially due to dyspnea and fatigue, and that avoidance of these activities over time occurs in patients. [15] As is known, because of symptoms of PAH, physical activities, exercise capacities, and muscle strengths of the individuals decrease.[12,16] According to the results of a large survey study which included 326 patients with PAH and 129 caregivers (total n=455), 56% of the patients with PAH reported very severe restriction of their activities of daily living.[17] It was also shown that with the increase in the functional classes of patients, a decrease in the perceived performance of activities of daily living occurs.[17] This study reveals the utmost importance of daily living activities both for the patients, and their relatives. However with progression in functional classification, the changes occurring in upper extremity functions and related performances of activities of daily living in patients with PAH should be disclosed. Thus, the impact of disease severity on these findings should be clearly determined to guide preparation of a treatment program suitable for the individual patient. In this context, the aim of this study was to determine the effect of disease severity on upper extremity muscle strength, exercise capacity and activities of daily living in persons with PAH.

Abbreviations:

6MWD	*6-min Walk Distance*
6MPRT	*6-min Pegboard and Ring Test*
COPD	*Chronic Obstructive Pulmonary Disease*
MAS	*Milliken Activities of Daily Living Scale*
NYHA	*New York Heart Association*
PAH	*Pulmonary arterial hypertension*
WHO	*World Health Organization*

METHODS

Patients

Twenty-five clinically stable patients with the diagnosis of PAH, and NYHA class II (n=14), and III (n=11) aged between 18, and 78 years who were followed up in the outpatient clinic of Dokuz Eylül University were enrolled in the study.

The stable NYHA functional Stage II and III patients who had undergone right heart catheterization, with resting median pulmonary artery pressure of ≥ 25 mm Hg who were receiving drug therapy for the last 3 months were included in the study. Patients with marked restrictive or obstructive pulmonary disease, orthopedic disease which will effect exercise tests, severe ischemic heart disease, left heart failure or acute cor pulmonale were not included in the study. Approval for the study was obtained from the Noninvasive Research Ethics Committee of Dokuz Eylül University. Consent forms were obtained from individuals enrolled in the study, and they were assured that their personal rights were protected.

Method

Demographic, physical, and physiologic characteristics (age, height, weight, body mass index, and pulmonary arterial pressure) of the patients with PAH were recorded.

Six-Minute - Pegboard and Ring Test (6PBRT): It was used to evaluate exercise capacity of upper extremities. In this test 4 iron rods, and a total of 20 rings are placed on a pegboard. The patients are requested to place with their both hands to place the rings on the rods firstly from top to bottom, then vice versa (Figure 1a). At the end of six minutes, total number of rings placed were counted. Before and after the test heart rate, blood pressure, respiratory rate,

and dyspnea, and fatigue were evaluated based on Modified Borg Scale.[18]

Peripheral Muscle Test: Shoulder flexor, elbow extensor, and elbow flexors were evaluated using digital dynamometer (Commander™ Powertrack II™, JTECH Medical, Midvale, UT, USA), and handgrip strength with Jamar® hand dynamometer (Patterson Medical, Warrenville, IL, USA) . Based on the standards recommended by American Society of Hand Therapists. The patient is seated with shoulder in adduction, and neutral rotation, elbow at 90º flexion, forearm at midrotation, and resting on a support, and wrist at neutral position. (Figure 1b).[19] Other muscle tests were performed while the patient was laid at supine position Shoulder flexor muscle strength was measured while the shoulder at 90º flexion, and elbow at extension. While elbow extensor muscle strength was measured with the shoulder, and forearm at neutral position, elbow at 90º flexion. Elbow flexor muscle strength was measured while the shoulder at neutral position, elbow at 90º flexion, and forearm pointing upward (Figure 1c). Muscle tests at every region were repeated thrice for both sides, and data obtained were recorded in kg, and the highest value of the three measurements were used.[20,21]

Milliken Activities of Daily Living Scale (MAS): For the evaluation of the limitation of the activities, the MAS whihc is a self-report scale was used. For each item, "ability" and "necessity" are measured simultaneously. MAS scale consists of a total of 47 items including preparing meals, and eating (8 items), self-care (9 items), dressing oneself (8 items), manual manipulation of objects (9 items), house cleaning, and washing clothes (7 items), and other activities (6 items). For scoring the level of ability to perform each item, a 5-point scale, and for the level of necessity to perform these items, a 3-point scale were used. Total score can be at most 705 points. The validity and reliability of the MAS for Turkish population have been investigated.[22]

Six-minute Walk Test (6MWT): In this study, the standard 6MWT protocol recommended by American Thoracic Society guideline was used.[23]

Statistical Analysis

Statistical analyses were performed using IBM SPSS Statistics (version 20) program. The variables were assessed for normality using the Shapiro-Wilk test, and investigating the histograms and probability graphs. Since the variables had non-normal distribution, the non-parametric test statistics were used. Continuous variables were expressed as median (interquartile range), while categorical variables were indicated as distribution of frequencies.

The significance of intergroup difference as for continuous variables with non-normal distribution was evaluated using Mann-Whitney U test. Intergroup difference (if any) was assessed as for gender, and clinical classification of PAH using Fisher test. Level of statistical significance was accepted as $p<0.05$.[2425]

RESULTS

Median ages (interquartile range) of the study participants were 61.5 (48.75-67.5), and 66.0 (53.0-71.0) years in groups of NYHA II, and III, respectively. Any intergroup difference was not found regarding age, gender, height, body weight, body mass index, mean pulmonary artery pressure, and characteristics of PAH clinical classification ($p>0.05$) (Table 1). Median (interquartile range) pulmonary artery pressures of the patients in groups of NYHA II, and III were 75.0 (65.75-103.75) mm Hg, and 90.0 (50.0-115.0) mm Hg, respectively. Patients in group NYHA II had idiopathic (n=10), and related to congenital heart diseases (n=4). The patients in the NYHA III group had idiopathic PAH

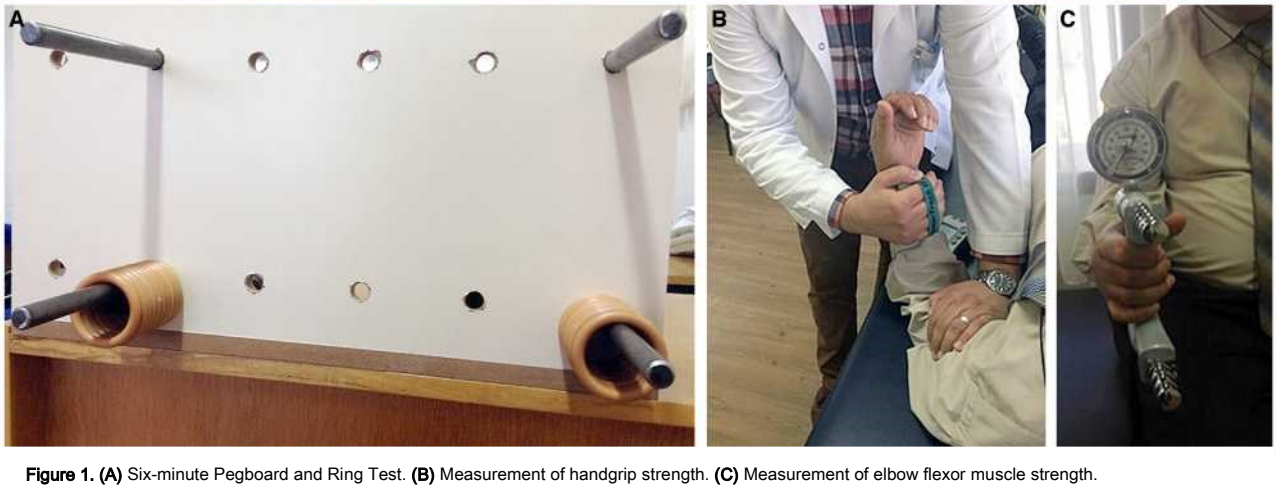

Figure 1. (A) Six-minute Pegboard and Ring Test. **(B)** Measurement of handgrip strength. **(C)** Measurement of elbow flexor muscle strength.

Table 1. Characteristic features of the individuals and their comparative analysis

	NYHA II (n=14)	NYHA III (n=11)	*p*
Age (years)	61.5 (48.75-67.5)	66.0 (53.0-71.0)	0.411
Gender, n (%)			
Female	10 (71.4)	10 (90.9)	0.341
Male	4 (28.6)	1 (9.1)	
Height (m)	1.60 (1.55-1.67)	1.60 (1.55-1.65)	0.691
Body weight (kg)	72.0 (62.25-81.87)	72.0 (59.0-80.0)	0.936
Body mass index (kg/m^2)	25.51 (22.64-33.29)	23.57 (23.04-23.57)	0.558
Clinical classification of PAH, n (%)			
Idiopathic PAH	10 (71.4)	6 (54.5)	0.434
PAH related with other diseases	4 (28.6)	5 (45.5)	
Median PAP (mmHg)	75.0 (65.75-103.75)	90.0 (50.0-115.0)	0.966

Continuous variables were expressed as median (interquartile range), and categorical variables as numbers and percentages. NYHA: New York Heart Association; PAP: Pulmonary artery pressure

Table 2. Clinical characteristics of the individuals and their comparative analysis

	NYHA II (n=14)	NYHA III (n=11)	*p*
6PBRT	151.0 (136.5–192.5)	115.0 (85–131)	0.004
MAS	626.0 (537–670)	429.5 (376.25–527.25)	0.002
Shoulder flexion–right (kg)	11.09 (8.32–12.28)	7.81 (5.15–10.82)	0.069
Shoulder flexion–left (kg)	9.96 (6.85–12.45)	8.15 (6.05–11.08)	0.363
Elbow flexion–right (kg)	11.55 (8.83–15.17	7.36 (5.15–10.64)	0.043
Elbow flexion–left (kg)	11.32 (6.69–14.60)	8.04 (6.28–11.78)	0.265
Elbow extension–right (kg)	8.83 (6.22–10.24)	6.56 (4.92–11.43)	0.538
Elbow extension–left (kg)	9.06 (5.92–10.70)	6.90 (4.86–10.30) 0	0.379
Grip strength–right (kg)	23.0 (21.5–35.5)	14.0 (11.75–18.75)	0.002
Grip strength–left (kg)	21.0 (19.0–37.5)	13.0 (10.0–17.25)	0.003
6MWD (m)	445 (390–490)	225 (180–280)	<0.001

Continuous variables were expressed as median (interquartile range), and categorical variables as numbers, and percentages. NYHA: New York Heart Association; 6PBRT: 6-Minute Pegboard and Ring Test; MAS: Milliken Activities of Daily Living Scale; 6MWD: 6-min walk Distance.

(n=6), and related to congenital heart diseases (n=3) and connective tissue diseases (n=2).

Clinical characteristics of PAH patients, and relevant comparisons according to NYHA II, and NYHA III are given in Table 2. Upper extremity exercise capacity, and MAS score were significantly lower in NYHA III group relative to NYHA II group (p=0.004 and p=0.002, respectively). In the NYHA III group elbow flexion (right), and handgrip strength (right, and left) were significantly lower relative to NYHA II group (p=0.043, p=0.002, and p=0.003, respectively). Besides in the NYHA III group 6MWD was significantly lower relative to NYHA II group (p<0.001). Strengths of shoulder flexor (right, and left), elbow flexor (left), and extensor (right, and left) muscles in NYHA class III were lower than the muscle strengths of NYHA class II patients, but a statistically significant intergroup difference was not seen (p>0.05).

DISCUSSION

This study showed that when the patients with PAH with different disease severity were compared, the patients with the NYHA functional class III had lower upper extremity exercise capacity, lower muscle strength of some upper extremities, and higher restrictions in activities of daily living related with upper extremities compared to the patients with functional class II. No significant difference between demographic characteristics of the patients including age, gender, and body mass index demonstrates that homogenous groups were compared.

In this study, it was shown that the upper extremity exercise capacity decreased as disease severity increased. Evaluation of exercise capacity of upper extremities, and activities of daily living in COPD patients are important for the assessment of the results of pulmonary rehabilitation.[26] In patients who received strengthening exercises of upper extremities, the 6PBRT has been demonstrated to be a test which can display the results of pulmonary rehabilitation program.[27] Besides, as reported in many studies the 6PBRT reflects the exercise capacity of upper extremity functions in activities of daily living.[28] However, since lower extremity exercise treatments are used more frequently in patients with PAH, the 6MWD is frequently used as an outcome measure.[29,30] Two studies evaluating the exercise capacity using the 6MWD showed that functional status was worsened as disease severity increased when compared to the World Health Organization (WHO) and NYHA classifications. [12,31] The importance of the evaluation of upper extremities, and its place in pulmonary rehabilitation increase day by day, and in the most updated current guidelines on pulmonary rehabilitation, the importance of the evaluation, and treatment of upper extremities were emphasized. [32] Based on the results of this study, in patients with PAH, in parallel with increase in the disease severity, decrease in the performance of upper extremity has been suggested. Based on the outcomes of these studies, in the evaluation, and treatment of the patients with PAH, upper extremities should be definitively taken into consideration.

In the NYHA III group, flexors of the right elbow, and grip strenghts of the right, and left arms, were demonstrated to be significantly lower when compared with the NYHA II group. In previous studies, hand grip strengths of the patients with PAH were demonstrated to be lower when compared with those of the healthy controls.[7] Patients with PAH who were categorized according to the WHO classification, and significantly decreased hand grip strengths in WHO II and WHO III groups were reported when compared with healthy controls.[12] In the only study in the literature where muscle strengths of upper extremities were compared, shoulder abductor muscle strengths of the patients in the groups WHO III, and II were found to be significantly decreased relative to the healthy group. [12] However, no significant difference between WHO III, and II as for shoulder abductor muscle strengths was demonstrated.[12] In our study, shoulder abductor muscle strengths were not measured, however when shoulder flexor strengths were compared, any intergroup difference was not found as detected in the previous studies. Similarly, even though any intergroup difference did not exist as for elbow extensor muscle strengths, it was lower in the NYHA III group. These results suggest that in patients with PAH, muscle strengths of especially distal upper extremities decrease, as the disease severity increases.

In this study, it has been demonstrated that restriction of activities of daily living involving upper extremities worsens, in parallel with increasing disease severity in patients with PAH. Similarly, it has been also reported that patients with COPD were compelled to decrease activities involving upper extremities as cooking a meal, and brushing teeth because of dyspnea, and arm fatigue.[33] Refraining from activities of daily living with increasing disease severity has been thought to be possibly related to decreased exercise capacity of the upper extremities.

Limitations

This study has some limitations. This study had a smaller sample size because of rarity of PAH. Besides, all groups included in the functional classification could be compared with each other. Since a homogenous distribution could not be achieved in NYHA I and NYHA IV classes, other groups were not included in the study. However comparison of other groups in larger scale studies will yield useful outcomes. Additionally, lack of age, and gender-matched healthy controls is one of the other limitations of the study. Inclusion of healthy individuals might provide more explanatory information about the extent of impact on the analyzed variables in the patients with PAH. Although reliability of the 6PBRT in patients with COPD was reported previously,[27] this test has not been applied in patients with PAH. However, this test, which is easier to use in the clinical practive, was preferred because of similar affected muscle strength and exercise capacity in the patients with PAH and COPD. The applicability of this test in patients with PAH should be further investigated.

In conclusion, in PAH patients, as disease severity increases, exercise capacity of upper extremities, strength of elbow flexors (right), and hand grip strength decrease, and restrictions in activities of daily living related with upper extremities increase. While rehabilitation programs are planning in patients with PAH, the disease severity should be considered and the evaluations and treatments for the upper extremities should be included.

Conflict of Interest: None declared

REFERENCES

1. Hoeper MM, Bogaard HJ, Condliffe R, Frantz R, Khanna D, Kurzyna M, et al. Definitions and diagnosis of pulmonary hypertension. J Am Coll Cardiol 2013;62:D42-50.
2. Galie N, Humbert M, Vachiery JL, Gibbs S, Lang I, Torbicki A, et al. 2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the Euro- pean Respiratory Society (ERS): Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). Eur Heart J 2016;37:67-119.
3. Halank M, Einsle F, Lehman S, Bremer H, Ewert R, Wilkens H, et al. Exercise capacity affects quality of life in patients with pulmonary hypertension. Lung 2013;191:337-43.
4. Fowler RM, Gain KR, Gabbay E. Exercise intolerance in pulmonary arterial hypertension. Pulm Med 2012;2012:359204.
5. Babu AS, Arena R, Myers J, Padmakumar R, Maiya AG, Cahalin LP, et al. Exercise intolerance in pulmonary hypertension: mechanism, evaluation and clinical implications. Expert Rev Respir Med 2016;10:979-90.
6. de Man FS, van Hees HW, Handoko ML, Niessen HW, Schalij I, Humbert M, et al. Diaphragm muscle fiber weak- ness in pulmonary hypertension. Am J Respir Crit Care Med 2011;183:1411-8.
7. Bauer R, Dehnert C, Schoene P, Filusch A, Bartsch P, Borst MM, et al. Skeletal muscle dysfunction in patients with idiopathic pulmonary arterial hypertension. Respir Med 2007;101:2366-9.
8. Tolle J, Waxman A, Systrom D. Impaired systemic oxygen extraction at maximum exercise in pulmonary hypertension. Med Sci Sports Exerc 2008;40:3-8.
9. Breda AP, Pereira de Albuquerque AL, Jardim C, Morinaga LK, Suesada MM, Fernandes CJ, et al. Skeletal muscle ab- normalities in pulmonary arterial hypertension. PLoS One 2014;9:e114101.
10. Sun XG, Hansen JE, Oudiz RJ, Wasserman K. Exercise pathophysiology in patients with primary pulmonary hypertension. Circulation 2001;104:429-35.
11. Miyamoto S, Nagaya N, Satoh T, Kyotani S, Sakamaki F, Fujita M, et al. Clinical correlates and prognostic significance of six-minute walk test in patients with primary pulmonary hypertension. Comparison with cardiopulmonary exercise testing. Am J Respir Crit Care Med 2000;161:487-92.
12. Saglam M, Vardar-Yagli N, Calik-Kutukcu E, Arikan H, Savci S, Inal-Ince D, et al. Functional exercise capacity, physical activity, and respiratory and peripheral muscle strength in pulmonary hypertension according to disease severity. J Phys Ther Sci 2015;27:1309-12.
13. McLaughlin VV, Archer SL, Badesch DB, Barst RJ, Farber HW, Lindner JR, et al; ACCF/AHA. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart As- sociation: developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. Circulation 2009;119:2250-94.
14. Pedretti LW, Pendleton HM, Schultz-Krohn W. Pedretti's occupational therapy : practice skills for physical dysfunction. Elsevier; 2013.
15. Calik-Kutukcu E, Arikan H, Saglam M, Vardar-Yagli N, Oksuz C, Inal-Ince D, et al. Arm strength training improves activities of daily living and occupational performance in patients with COPD. Clin Respir J 2015 Dec 1 [Epub ahead of print], doi: 10.1111/crj.12422.
16. Okumus G, Aslan GK, Arseven O, Ongen G, Issever H, Kıyan E. The role of an activity monitor in the objective evaluation of patients with pulmonary hypertension. Clin Respir J 2016 May 5 [Epub ahead of print], doi: 10.1111/crj.12495.
17. Guillevin L, Armstrong I, Aldrighetti R, Howard LS, Ryftenius H, Fischer A, et al. Understanding the impact of pulmonary arterial hypertension on patients' and carers' lives. Eur Respir Rev 2013;22:535-42.
18. Celli BR, Rassulo J, Make BJ. Dyssynchronous breathing during arm but not leg exercise in patients with chronic air- flow obstruction. N Engl J Med 1986;314:1485-90.
19. Haidar SG, Kumar D, Bassi RS, Deshmukh SC. Average versus maximum grip strength: which is more consistent? J Hand Surg Br 2004;29:82-4.
20. Bohannon RW. Reference values for extremity muscle strength obtained by hand-held dynamometry from adults aged 20 to 79 years. Arch Phys Med Rehabil 1997;78:26-32.
21. Andrews AW, Thomas MW, Bohannon RW. Normative values for isometric muscle force measurements obtained with hand-held dynamometers. Phys Ther 1996;76:248-59.
22. Akel BS, Oksuz C, Kayihan H. Adaptation and validation of Turkish version of the Milliken ADL Scale (MAS). J Hand Ther 2010;23:e12.
23. ATS Committee on Proficiency Standards for Clinical Pulmonary Function Laboratories. ATS statement: guidelines for the six-minute walk test. Am J Respir Crit Care Med 2002;166:111-7.
24. Hayran M, Hayran M. Sağlık araştırmaları için temel istatistik. Ankara: Omega Araştırma; 2011.
25. Ortiz-Gutierrez R, Cano-de-la-Cuerda R, Galan-del-Rıo F, Alguacil-Diego IM, Palacios-Cena D, Miangolarra-Page JC. A telerehabilitation program improves postural control in multiple sclerosis patients: a Spanish preliminary study. Int J Environ Res Public Health 2013;10:5697-710.
26. Takeda K, Kawasaki Y, Yoshida K, Nishida Y, Harada T, Yamaguchi K, et al. The 6-minute pegboard and ring test is correlated with upper extremity activity of daily living in chronic obstructive pulmonary disease. Int J Chron Obstruct Pulmon Dis 2013;8:347-51.
27. Janaudis-Ferreira T, Hill K, Goldstein RS, Wadell K, Brooks D. Relationship and responsiveness of three upper-limb tests in patients with chronic obstructive pulmonary disease. Physiother Can 2013;65:40-3.

28. Janaudis-Ferreira T, Beauchamp MK, Goldstein RS, Brooks D. How should we measure arm exercise capacity in patients with COPD? A systematic review. Chest 2012;141:111-20.
29. Sanchis-Gomar F, Gonzalez-Saiz L, Sanz-Ayan P, Fiuza-Lu- ces C, Quezada-Loaiza CA, Flox-Camacho A, et al. Ratio- nale and Design of a Randomized Controlled Trial Evaluating Whole Muscle Exercise Training Effects in Outpatients with Pulmonary Arterial Hypertension (WHOLEi+12). Cardiovasc Drugs Ther 2015;29:543-50.
30. Bussotti M, Gremigni P, Pedretti RF, Kransinska P, Di Marco S, Corbo P, et al. Effects of an outpatient service rehabilitation programme in patients affected by pulmonary arterial hypertension: an observational study. Cardiovasc Hematol Disord Drug Targets. 2016 Nov 30 [Epub ahead of print].
31. Miyamoto S, Nagaya N, Satoh T, Kyotani S, Sakamaki F, Fujita M, et al. Clinical correlates and prognostic significance of six-minute walk test in patients with primary pulmonary hypertension. Comparison with cardiopulmonary exercise testing. Am J Respir Crit Care Med 2000;161:487-92.
32. Spruit MA, Singh SJ, Garvey C, ZuWallack R, Nici L, Rochester C, et al. An official American Thoracic Society/European Respiratory Society statement: key concepts and advanc- es in pulmonary rehabilitation. Am J Respir Crit Care Med 2013;188:e13-64.
33. Pan L, Guo YZ, Yan JH, Zhang WX, Sun J, Li BW. Does upper extremity exercise improve dyspnea in patients with COPD? A meta-analysis. Respir Med 2012;106:1517-25.

Anahtar sözcükler: Pulmoner arteriyel hipertansiyon; üst ekstremite egzersiz kapasitesi; üst ekstremite günlük yaşam aktiviteleri; üst ekstremite kas kuvveti.
Keywords: Pulmonary arterial hypertension upper extremity exercise capacity; upper extremity activities of daily living; upper extremity muscle strength.