

Muscle Strength and Activation as Determinants of Upper Extremity Exercise Capacity in Adolescent and Young Adult Patients with Pulmonary Arterial Hypertension

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ABSTRACT

Purpose: Pulmonary arterial hypertension (PAH) impairs aerobic capacity due to combined cardiopulmonary and skeletal muscle dysfunction. However, upper extremity aerobic performance and its muscle-related correlates remain underexplored. This study aimed to examine the relationship between maximal workload achieved during an arm ergometer test and peripheral muscle parameters in adolescents and young adults with PAH.

Method: Sixteen PAH patients (mean age: 19.88 ± 7.44 years; 9 females) were assessed. Maximal workload (watts) was measured via arm ergometry. Handgrip and shoulder abduction strength were recorded using dynamometers and normalized to percent predicted values. Fat-free mass (FFM) was estimated via bioelectrical impedance analysis. Surface electromyography (EMG) of the anterior deltoid was collected during the initial and final stages of the exercise and expressed as %Maximum Voluntary Isometric Contraction (MVIC) following SENIAM standards. Brain Natriuretic Peptide (BNP) values were retrieved from clinical records. Spearman correlation was used to assess associations.

Results: Maximal workload showed positive correlations with FFM ($r=0.615$, $p=0.011$), shoulder abduction strength ($r=0.535$, $p=0.033$), and handgrip strength ($r=0.620$, $p=0.010$). It was negatively correlated with BNP ($r=-0.728$, $p=0.005$) and early-stage anterior deltoid EMG (%MVIC) ($r=-0.602$, $p=0.014$). Final-stage EMG showed a non-significant negative trend ($r=-0.442$, $p=0.086$).

Conclusion: Upper extremity aerobic performance in PAH is associated with muscle strength, mass, activation, and cardiac stress. Arm ergometry may serve as a functional indicator in this population.

Key Words: Upper Extremity, Motor Activity, Electromyography, Muscle Strength, Pulmonary Hypertension

INTRODUCTION

Pulmonary hypertension (PH) is defined as a mean pulmonary arterial pressure exceeding 20 mmHg at rest. Despite advancements in medical management, patients with PH often experience persistent symptoms that negatively affect their quality of life. Among these, exercise intolerance is a predominant complaint and has been widely recognized as a critical prognostic indicator. However, the mechanisms underlying reduced exercise capacity in PH remain only

partially understood, making them a key target for novel assessment and intervention strategies (1).

Pulmonary arterial hypertension (PAH) is a rare, progressive cardiopulmonary disease characterized by increased pulmonary vascular resistance and right ventricular dysfunction, leading to reduced oxygen delivery and exercise intolerance (2). In both adults and pediatric populations, this intolerance is not solely a consequence of cardiopulmonary impairment; it's also heavily influenced by peripheral skeletal

muscle dysfunction. Recent studies have demonstrated that muscle abnormalities such as reduced muscle mass, decreased oxidative capacity, fiber type shifting from type I to type II, and diminished capillarization play critical roles in limiting functional performance in PAH patients (3).

Skeletal muscle weakness in PAH has been well documented, particularly in the lower limbs, but emerging evidence also indicates clinically significant impairments in upper extremity strength. This muscle weakness is driven by chronic hypoxia-induced mitochondrial dysfunction, impaired protein synthesis, and inflammatory cytokine activation, which collectively reduce contractile efficiency and oxidative metabolism (4). Decreased muscle strength has been associated with reduced peripheral oxygen extraction, lower mitochondrial density, and impaired metabolic efficiency. These alterations not only affect functional endurance but also contribute to higher perception of fatigue during daily activities.

While lower extremity performance has been extensively studied in PAH, upper extremity function remains an underexplored but clinically significant area. Upper extremity muscles are crucial for daily life activities such as eating, hygiene, and dressing. Previous research by Camcıoğlu (2017), has shown that upper extremity aerobic exercise training improves functional capacity, respiratory and peripheral muscle strength, and quality of life in PAH patients. However, there is a gap in the literature regarding the physiological and neuromuscular responses observed during acute upper limb aerobic effort, particularly the interplay between muscle strength, muscle mass, and muscle activation (5).

Surface electromyography (sEMG), especially when normalized to maximum voluntary isometric contraction (MVIC), is a reliable method for assessing neuromuscular efficiency and muscle recruitment patterns during exercise (6). sEMG can help clarify how efficiently muscles are activated in response to submaximal workloads and how this relates to fatigue and exercise limitation in clinical populations, including PAH. To date, no studies have directly examined EMG-derived activation patterns of upper extremity muscles such as the anterior deltoid during aerobic

exercise in patients with PAH, highlighting a gap in the current literature. Moreover, biomarkers such as B-type natriuretic peptide (BNP), which reflect cardiac stress, have been correlated with reduced exercise capacity and increased symptom severity in PAH (7, 8). Integrating muscular and cardiac parameters may provide a more comprehensive understanding of the limitations faced by PAH patients during physical activity

Understanding these interrelations can help identify peripheral contributors to exercise limitation and inform tailored rehabilitation approaches that incorporate muscle-targeted interventions alongside cardiopulmonary care. Therefore, the aim of this study was to investigate the associations between maximum workload achieved during an upper extremity aerobic exercise test and peripheral muscle strength (handgrip and shoulder abduction), muscle mass (fat-free mass), and anterior deltoid muscle activation (via sEMG), as well as BNP levels, in adolescents and young adults diagnosed with PAH.

METHODS

Study Design and Participants

This cross-sectional observational study included adolescents and young adults diagnosed with World Health Organization (WHO) Group 1 PAH. Participants were recruited from the Hacettepe University Pediatric and Adult Pulmonary Hypertension Clinics. Participant recruitment took place between January and July 2024. Participants were eligible for inclusion if they were between 10 and 30 years of age, had a confirmed diagnosed PAH, and were clinically stable to complete exercise testing procedures. Individuals were excluded if they had any neuromuscular or orthopedic condition affecting upper extremity function, cognitive or behavioral impairments interfering with test instructions, or incomplete physiological recordings that compromised data quality. Two individuals were subsequently excluded from final analyses due to incomplete or invalid testing data, resulting in a sample of 16 participants (mean age 19.88 ± 7.44 years; 9 females). All participants and/or their legal guardians provided written informed consent. Ethical approval for the study was granted by the Hacettepe University Non-

Interventional Clinical Research Ethics Committee (Approval No: GO 22/39), and the study was conducted in accordance with the Declaration of Helsinki.

Testing Protocol and Outcome Measures

All evaluations were carried out in a controlled laboratory setting in a single session at the Pediatric Cardiology Department and the Faculty of Physical Therapy and Rehabilitation of Hacettepe University. Participants were advised to avoid vigorous physical activity, caffeine, and heavy meals for 12 hours prior to testing. At study entry, basic clinical and demographic information was obtained through medical record review and participant (or parent) interview. Data collected included age, sex, height, weight, disease duration, diagnosis, current medications, and BNP levels. The assessment battery included measurements of body composition, upper body aerobic performance, muscle strength and sEMG.

New York Heart Association (NYHA) Functional Classification Assessment

Patients' functional status was assessed using the NYHA functional classification system, which stratifies individuals into four categories based on the severity of symptoms experienced during physical activity. Classification was conducted during the initial clinical evaluation through a structured interview. Participants, and caregivers when applicable, were asked about the presence and severity of exertional dyspnea, fatigue, and limitations in performing age-appropriate daily activities. In pediatric and adolescent populations, this approach has been validated and adapted for use in research and clinical settings (9).

1. Upper Extremity Aerobic Test (Arm Ergometer Protocol): Upper extremity aerobic capacity was assessed using a graded submaximal arm ergometer protocol (Monark 891E, Sweden). Participants were seated in a height-adjustable chair with back support, and the ergometer crank was aligned with shoulder level. After a 5-minute rest, a 2-minute warm-up was performed at 0 W and 60–75 rpm. The test consisted of 2-minute stages with workload increased by 10 W at each stage. The test was terminated upon volitional

exhaustion, inability to maintain rpm, or occurrence of predefined clinical termination criteria (e.g., systolic blood pressure >220 mmHg, oxygen saturation (SpO₂) drop ≥15%, or a modified Borg Scale score ≥8). Throughout the test, heart rate (HR) and peripheral oxygen saturation (SpO₂) were continuously monitored, while dyspnea and arm fatigue were assessed using the modified Borg CR-10 scale. The primary aerobic performance outcome was the maximum workload (Watts) reached during the test.

2. Muscle Strength Measurement: Handgrip strength was measured using a calibrated Jamar hydraulic hand dynamometer (Model J00105, Patterson Medical, USA). Participants were seated with the elbow flexed at 90°, the forearm in a neutral position, and the wrist in slight extension, following standardized testing protocols. Three maximal isometric contractions were performed with the dominant hand, with a 60-second rest between trials. The highest value (in kilogramforce) was recorded and used for analysis (10).

Shoulder abduction strength was measured using a Lafayette digital handheld dynamometer (Model 01165, USA). Participants performed isometric contractions against resistance at 90° shoulder abduction while seated (11). Shoulder abduction strength values were converted into percent predicted (% predicted) based on age- and sex-adjusted normative data (12, 13).

3. Body Composition Analysis: Body composition was measured using a bioelectrical impedance analyzer (Xiaomi Mi Body Composition Scale 2, China) (14). During the measurement, participants were asked to step on the scale with light clothing and bare feet and remain motionless for a short time. The data obtained from the scale was transferred via the phone application and analyzed to obtain body composition results including fat-free mass and body fat percentage (15).

4. Electromyographic Muscle Activation: sEMG was used to assess neuromuscular activation of the anterior deltoid muscle in the dominant arm. Disposable Ag/AgCl electrodes were placed following the SENIAM (Surface EMG for Non-Invasive Assessment of Muscles) guidelines. Prior to electrode placement, the skin was shaved and cleaned with

alcohol to reduce impedance. sEMG signals were collected using a Noraxon DTS system (Noraxon USA Inc.) with a sampling rate of 1000 Hz and a band-pass filter of 20–450 Hz.

During the arm ergometer aerobic test, muscle activation was analyzed from two specific time windows: the first 15 seconds of the initial workload stage and the final 15 seconds of the last tolerated workload stage. These segments were selected to represent early and late phase activation. The 15-second time windows were selected to reduce signal noise and standardize the comparison across participants by minimizing variability due to transient fluctuations. All EMG signals were normalized to MVIC, which was measured via a standardized isometric contraction at 90° shoulder flexion. Figure 1 presents individual level sEMG data, illustrating changes in anterior deltoid activation between the first and final workload stages of the upper extremity exercise test. EMG values were expressed as %MVIC (6, 7).

Sample Size Determination

A preliminary pilot analysis involving five participants was conducted to examine the associations between upper extremity muscle strength and activation and aerobic performance, represented by maximal workload (W_{max}) during the arm ergometer test. Key variables included handgrip strength, shoulder abduction strength, and anterior deltoid sEMG. Correlation analyses were performed using SPSS v.25. Based on the effect sizes observed in the pilot ($r = 0.50$ – 0.70), a power analysis with G*Power 3.1 indicated that a sample size of 16 would be sufficient to detect moderate-to-strong associations ($r \geq 0.60$) with a power of 0.80 and $\alpha = 0.05$ (two-tailed).

Statistical Analysis

All statistical analyses were performed using IBM SPSS Statistics version 25.0 (IBM Corp., Armonk, NY, USA). Descriptive statistics were presented as mean \pm standard deviation (SD) or median (interquartile range, IQR) based on normality. The Shapiro–Wilk test was used to assess normality. Because several variables violated the assumption of normal distribution, Spearman's rank correlation coefficient (r) was used to evaluate associations between

maximum workload (Watts) and muscle strength, body composition, EMG muscle activity, and BNP. Statistical significance was set at $p < 0.05$. The strength of correlations was interpreted using standard thresholds: negligible ($r < 0.20$), weak ($r = 0.20$ – 0.39), moderate ($r = 0.40$ – 0.59), strong ($r = 0.60$ – 0.79), and very strong ($r \geq 0.80$) (16).

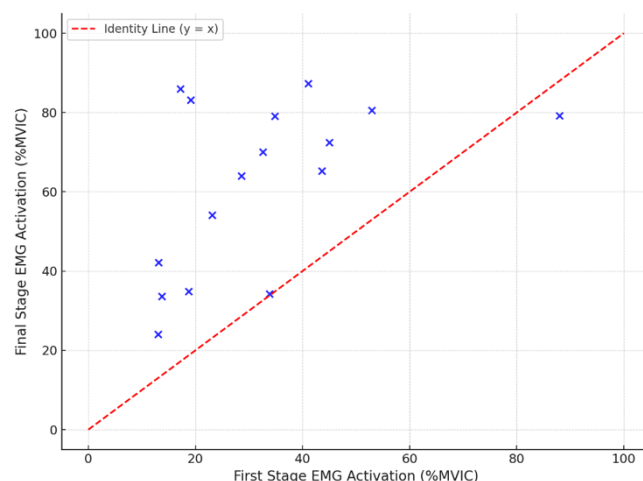


Figure 1. Scatter plot illustrating individual changes in sEMG activation of the right anterior deltoid during the upper extremity incremental exercise test. Each point represents one participant's activation level (%MVIC) during the first and final tolerated workload stages. The dashed identity line ($y = x$) indicates equal activation between stages. Most data points appear above the line, reflecting increased neuromuscular demand toward the end of the test.

RESULTS

A total of 16 participants with a diagnosis of PAH (9 females, 7 males) were included in the analysis. The mean age was 19.88 ± 7.44 years, and the mean body mass index (BMI) was 18.97 ± 6.63 kg/m². The mean FFM was 41.82 ± 15.61 kg, and the mean body fat percentage was $12.76 \pm 9.58\%$. Based on NYHA functional classification, 18.8% were Class I, 56.3% Class II, and 25% Class III. The median disease duration was 19.5 years (range: 9–30), mean ejection fraction was $63.16 \pm 11.14\%$, and mean pulmonary artery pressure was 61.75 ± 29.75 mmHg (Table 1).

During the arm ergometer aerobic test, participants achieved a median maximum workload of 40 watts (range: 20–50). HR, systolic and diastolic blood pressures increased significantly post-test, while SpO₂ declined by $-5.93 \pm 3.62\%$. Dyspnea and perceived arm fatigue scores increased markedly. Shoulder abduction strength averaged 12.85 ± 6.18 kg ($71.58 \pm 36.60\%$ of predicted), and hand grip strength was

24.12±10.15 kg. EMG-derived anterior deltoid activation was 39.26±36.32 %MVIC at the beginning and 61.87±21.59 %MVIC at the final stage of the test (Table 2).

Table 1. Demographic and Clinical Parameters

Variables		PAH (n=16)
Gender	Male	7 (% 43.80)
	Female	9 (% 56.2)
Age (years)		19.88 ± 7.44
BMI (kg/m ²)		18.97± 6.63
FFM (kg)		41.82±15.61
Body Fat (%)		12.76±9.58
NYHA Functional Class		
- Class I		3 (% 18.8)
- Class II		9 (% 56.30)
- Class III		4 (% 25)
Disease Duration (years)		19.50 (9-30)
Ejection Fraction (%)		63.16±11.14
Mean Pulmonary Artery Pressure (mmHg)		61.75±29.75
BNP (pg/mL)		122.9 (7.51 – 1208)
Diagnosis (n/%): Eisenmenger (6, 33.3%), Primary PAH (4, 22.2%), Residual PAH (3, 16.7%), Fontan (2, 11.1%), Glenn (1, 5.6%)		

Data are presented as mean ± standard deviation (SD), median (range), or frequency (n, %), as appropriate. BMI: Body Mass Index; FFM: Fat-Free Mass; NYHA: New York Heart Association Functional Class.

Spearman correlation analysis showed that FFM was strongly positively correlated with maximum workload ($r=0.615$, $p=0.011$), as was hand grip strength ($r=0.620$, $p=0.010$). Shoulder abduction strength showed a moderate positive correlation ($r=0.535$, $p=0.033$) These findings are detailed in Table 3. Post-test SpO₂ was moderately positively associated with workload ($r=0.579$, $p=0.019$), while Δ SpO₂ (the change from pre- to post-test) showed no association ($r=0.017$, $p=0.952$). BNP levels demonstrated a strong negative correlation with workload ($r=-0.728$, $p=0.005$). Maximum deltoid muscle activation at the beginning of the test was

moderate-to-strong negatively correlated with workload ($r=-0.602$, $p=0.014$), end-stage activation exhibited a weak negative trend that did not reach statistical significance ($r=-0.442$, $p=0.086$). These results are presented in Figure 2 and Table 3.

DISCUSSION

The primary aim of this study was to examine the relationship between upper extremity aerobic performance and peripheral muscle parameters in adolescents and young adults with PAH. To our knowledge, this is the first study to investigate how indicators of muscle mass, strength, and activation specifically in the upper limbs relate to functional aerobic capacity in this population using a standardized arm ergometer protocol. Our key finding was that the maximum workload achieved during the test was significantly associated with FFM, handgrip strength, shoulder abduction strength, and anterior deltoid muscle activation recorded via sEMG and normalized to maximum %MVIC. These results highlight the critical role of skeletal muscle health in determining exercise performance in PAH, an aspect that may be under-recognized in clinical settings traditionally focused on central hemodynamics.

Previous studies have consistently reported skeletal muscle dysfunction in PAH, particularly in the lower extremities, with reductions in muscle strength, oxidative capacity, and endurance (4, 17). The findings from the present study expand this knowledge by demonstrating that upper extremity muscle strength and mass also contribute significantly to aerobic performance. Our results align with the study by de Man et al., which showed improvements in quadriceps strength and endurance following exercise training in Idiopathic Pulmonary Arterial Hypertension (iPAH) patients (18). This supports the notion that muscle status is a modifiable factor influencing functional capacity in PAH. While de Man et al. focused exclusively on the lower limbs, our findings confirm comparable associations for shoulder and handgrip strength, reinforcing the systemic nature of skeletal muscle impairment in this disease (18).

Table 2. Physiological Parameters, Perceived Exertion, Muscle Strength, and Deltoid EMG Activation during the Arm Ergometer Test in PAH Patients

Parameter	Time Point	Value
Heart Rate (bpm)	Pre-test	88.93± 18.30
	Post-test	125.75±24.19
Systolic Blood Pressure (mmHg)	Pre-test	99.37± 10.93
	Post-test	122.81±11.96
Diastolic Blood Pressure (mmHg)	Pre-test	58.43± 11.50
	Post-test	68.43± 9.43
SpO ₂ (%)	Pre-test	87.31± 7.15
	Post-test	81.37± 7.70
ΔSpO ₂		-5.93± 3.62
Dyspnea Score (modified Borg Scale 0-10)	Pre-test	0.18± 0.75
	Post-test	3.06± 2.35
Arm Fatigue Score (modified Borg Scale 0-10)	Pre-test	2.87±2.47
	Post-test	7.18± 2.10
Shoulder Abduction Strength (kg)		12.85± 6.18
Shoulder Abduction Strength (% predicted)		71.58± 36.60
Hand Grip Strength (kg)		24.12± 10.15
Aerobic test max workload (watt)		40 (20-50)
Deltoid EMG – Max (%MVIC, final 15 s)	Beginning stage	39.26± 36.32
Deltoid EMG – Max (%MVIC, final 15 s)	End Stage	61.87± 21.59

This table presents pre- and post-test physiological responses (heart rate, blood pressure, oxygen saturation), perceived exertion scores (dyspnea and arm fatigue), upper extremity muscle strength values, and anterior deltoid surface EMG activity recorded during the arm ergometer aerobic test in patients with PAH. Data are expressed as mean ± standard deviation (SD) unless otherwise stated. ΔSpO₂ represents the change in peripheral oxygen saturation from pre-to-post-test. Abbreviations: SpO₂ = peripheral oxygen saturation; MVIC = maximal voluntary isometric contraction; EMG = electromyography; PAH = pulmonary arterial hypertension

Table 3: Correlations between Physiological and Muscle Parameters and Maximum Workload Achieved During Arm Ergometer Aerobic Test in PAH Patients

PAH (n=16)	Arm Ergometer Aerobic Test Maximum Workload Achieved (watts)	
	r	p
Fat Free Mass (kg)	0.615	0.011
Shoulder Abduction Strength (%)	0.535	0.033
Hand Grip Strength (kg)	0.620	0.010
Δ During Test SpO ₂ (%)	0.017	0.952
End Test SpO ₂ (%)	0.579	0.019
BNP (pg/mL)	-0.728	0.005
Arm Ergometer Test Maximum Muscle Activation at the Ending Stage (% MVIC)	-0.442	0.086
Arm Ergometer Test Maximum Muscle Activation at Beginning Stage (% MVIC)	-0.602	0.014

This table presents Spearman correlation coefficients (r) and corresponding p-values for the associations between maximum workload achieved during the arm ergometer aerobic test and physiological or muscle-related variables in patients with pulmonary arterial hypertension (PAH). Analyzed variables include fat-free mass (FFM), upper extremity muscle strength (shoulder abduction and handgrip), oxygen saturation (SpO₂), brain natriuretic peptide (BNP), and anterior deltoid muscle activation measured via surface EMG at the beginning and end stages of the test. Abbreviations: FFM = fat-free mass; SpO₂ = peripheral oxygen saturation; MVIC = maximal voluntary isometric contraction; BNP = brain natriuretic peptide; EMG = electromyography; PAH = pulmonary arterial hypertension.

Anterior deltoid muscle activation recorded via sEMG provided novel insight into peripheral muscle function during

upper extremity aerobic effort. In this study, EMG signals were collected during the first and final 15 seconds of the arm

ergometer test and normalized to %MVIC in accordance with SENIAM guidelines (6). Our findings showed that higher activation levels during the initial stage were moderate-to-strong and negatively correlated with maximum workload achieved, suggesting that participants with lower aerobic performance may require earlier and greater motor unit recruitment. Although activation measured at the final stage also showed a negative trend, this association did not reach statistical significance, likely reflecting inter-individual variability in neuromuscular fatigue onset. To our knowledge, no prior studies have quantified deltoid muscle activation using surface EMG during aerobic upper extremity testing in patients with PAH. While EMG methodology has been widely applied to respiratory muscles during exercise in cardiopulmonary populations, its use in upper limb skeletal muscles remains limited. Several studies have described similar EMG activation characteristics in respiratory muscles particularly the diaphragm and parasternal intercostals during exercise in clinical populations such as COPD, left heart failure, and PAH (19-21). These studies have shown increased early recruitment and higher EMG amplitudes relative to ventilatory or pressure output during submaximal tasks, suggesting altered neuromechanical coupling and reduced efficiency in respiratory muscle function (20). Our study extends EMG-based analysis to upper limb skeletal muscle during aerobic testing in PAH, offering a novel methodological perspective that may complement respiratory assessments in this population.

BNP levels demonstrated a strong negative correlation with maximum workload, indicating that elevated cardiac stress was associated with poorer aerobic performance. BNP is a well-established biomarker of right ventricular dysfunction and pulmonary vascular load in PAH, and its prognostic relevance has been confirmed in multiple clinical studies. For instance, Nagaya et al. showed that higher BNP levels were associated with increased pulmonary vascular resistance and decreased cardiac output, correlating inversely with peak oxygen uptake and 6-minute walk distance in PAH patients (22, 23). Similarly, Leuchte et al. reported that elevated NT-proBNP levels were inversely correlated with exercise

capacity and positively associated with pulmonary vascular resistance (8). Together, these findings suggest that central hemodynamic burden is a key contributing factor to reduced exercise capacity in patients with PAH

In contrast, post-test SpO₂ showed a moderate positive correlation with workload, implying that patients who were able to preserve systemic oxygenation during upper extremity exercise achieved better functional outcomes. Preserved oxygenation may reflect a combination of sufficient ventilatory reserve, maintained cardiac output, and efficient peripheral oxygen extraction. Interestingly, the change in SpO₂ during the test (Δ SpO₂) did not correlate with workload, suggesting that absolute oxygenation levels at peak exertion, rather than the magnitude of decline, may be more relevant in determining exercise performance. These findings emphasize the multifactorial nature of effort limitation in PAH, integrating cardiac, pulmonary, and peripheral components.

These findings underscore the importance of incorporating peripheral muscle assessment and targeted training into rehabilitation programs for patients with PAH. Given that muscle strength, neuromuscular activation patterns, and oxygenation were all significantly associated with aerobic performance in our study, a multimodal training approach that addresses both cardiovascular and skeletal muscle systems appears most promising. This perspective aligns with current rehabilitation guidelines for PAH, which recommend combining aerobic, resistance, and respiratory modalities to optimize patient outcomes. Prior clinical trials have demonstrated that such integrative interventions can lead to significant improvements in exercise capacity, WHO functional class, and quality of life (24-26).

Although previous studies have primarily focused on lower limb training, recent work in patients with COPD has shown that upper extremity resistance training can improve deltoid and trapezius activation, neuromuscular efficiency, and functional performance during daily tasks (21). These findings, together with our results, justify the inclusion of upper limb-specific exercises in PAH rehabilitation,

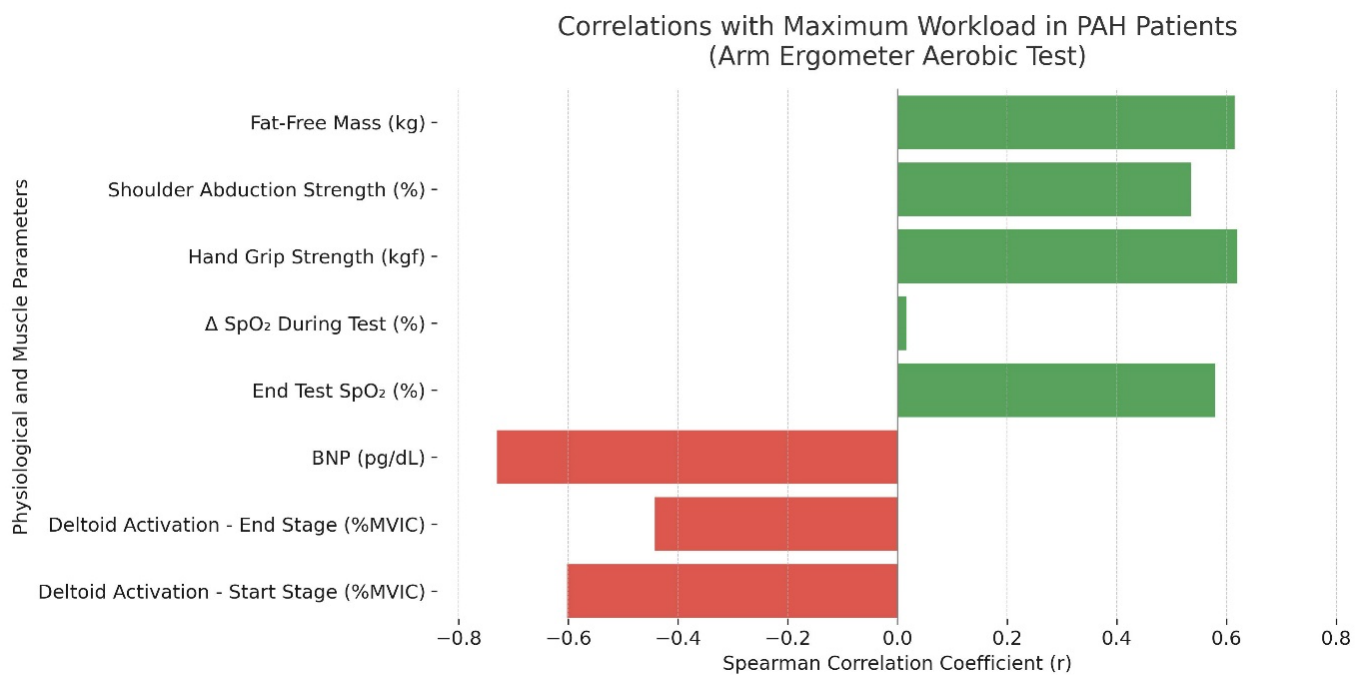


Figure 2. Correlations Between Physiological and Muscle Parameters and Maximum Workload in PAH Patients During the Arm Ergometer Aerobic Test. This figure presents Spearman correlation coefficients (r) and p -values for the relationship between the maximum workload achieved during the arm ergometer aerobic test and various physiological and muscle-related variables in patients with pulmonary arterial hypertension (PAH). Fat-free mass, upper extremity strength, oxygen saturation (SpO_2), and EMG-derived muscle activation parameters were analyzed. %MVIC indicates the percentage of maximum voluntary isometric contraction. BNP, B-type natriuretic peptide.

especially for younger patients in whom early neuromuscular intervention may slow the progression of physical deconditioning and improve long-term functional prognosis.

Several limitations of this study should be acknowledged. First, the relatively small sample size may have limited the statistical power to detect weaker associations or subgroup differences. Second, the cross-sectional design precludes causal interpretations; longitudinal studies are needed to determine whether interventions targeting muscle mass or neuromuscular function can lead to improvements in aerobic capacity. Third, although sEMG analysis offered useful insight into anterior deltoid activation patterns, it was restricted to a single superficial muscle. Broader assessments including multiple upper and lower limb muscles, as well as measures of muscular fatigue or recruitment dynamics, would provide a more comprehensive understanding of peripheral limitations. Finally, the absence of a healthy control group limits the ability to contextualize the degree of impairment observed. Future studies should include age and sex-matched healthy participants and assess the effects of multimodal training interventions on both central hemodynamics and peripheral muscle function

CONCLUSION

This study demonstrated that upper extremity muscle strength, mass, and activation patterns are significantly associated with aerobic performance in adolescents and young adults with PAH. Our findings highlight the importance of evaluating peripheral muscle function beyond cardiopulmonary parameters alone when assessing functional capacity in this population. These results support the integration of muscle-focused assessments and upper limb-specific exercise interventions into rehabilitation strategies for PAH, particularly in younger individuals. Further longitudinal research is warranted to determine the clinical impact of multimodal training approaches on long-term outcomes.

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REFERENCES

1. Vanhoof JM, Delcroix M, Vandeveld E, Denhaerynck K, Wuyts W, Belge C, et al. Emotional symptoms and quality of life in patients with pulmonary arterial hypertension. *The Journal of Heart and Lung Transplantation*. 2014;33(8):800-8.
2. Humbert M, Kovacs G, Hoeper MM, Badagliacca R, Berger RM, Brida M, et al. 2022 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension: Developed by the task force for the diagnosis and treatment of pulmonary hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS). Endorsed by the International Society for Heart and Lung Transplantation (ISHLT) and the European Reference Network on rare respiratory diseases (ERN-LUNG). *European heart journal*. 2022;43(38):3618-731.
3. Peng Zhang DDSGB, Alexander Vang, Julia Feord, Danielle J. McCullough, Alexsandra Zimmer, Natalie D'Silva, Richard T. Clements, Gaurav Choudhary. Reduced exercise capacity occurs before intrinsic skeletal muscle dysfunction in experimental rat models of pulmonary hypertension. *Pulmonary circulation*. 2024;14(2).
4. Mainguy V, Maltais F, Saey D, Gagnon P, Martel S, Simon M, et al. Peripheral muscle dysfunction in idiopathic pulmonary arterial hypertension. *Thorax*. 2010;65(2):113-7.
5. Burcu Camcıoğlu Yılmaz MBG, Müşerref Nur Keleş, Gülten Aydoğdu Taçoy, Atiye Çengel. Effects of upper extremity aerobic exercise training on oxygen consumption, exercise capacity, dyspnea and quality of life in patients with pulmonary arterial hypertension. *Heart & Lung*. 2020;49(5):564-71.
6. Hermens HJ, Freriks B, Disselhorst-Klug C, Rau G. Development of recommendations for SEMG sensors and sensor placement procedures. *J Electromyogr Kinesiol*. 2000;10(5):361-74.
7. Sturma A, Hruby LA, Prahm C, Mayer JA, Aszmann OC. Rehabilitation of Upper Extremity Nerve Injuries Using Surface EMG Biofeedback: Protocols for Clinical Application. *Frontiers in Neuroscience*. 2018;12.
8. Leuchte HH, Ten Freyhaus H, Gall H, Halank M, Hoepfer MM, Kaemmerer H, et al. Risk stratification strategy and assessment of disease progression in patients with pulmonary arterial hypertension: updated recommendations from the Cologne Consensus Conference 2018. *International journal of cardiology*. 2018;272:20-9.
9. Ivy DD, Abman SH, Barst RJ, Berger RM, Bonnet D, Fleming TR, et al. Pediatric pulmonary hypertension. *Journal of the American College of Cardiology*. 2013;62(25S):D117-D26.
10. Hamilton GF, McDonald C, Chenier TC. Measurement of grip strength: validity and reliability of the sphygmomanometer and jamar grip dynamometer. *Journal of Orthopaedic & Sports Physical Therapy*. 1992;16(5):215-9.
11. Mijnders DM, Meijers JM, Halfens RJ, ter Borg S, Luiking YC, Verlaan S, et al. Validity and reliability of tools to measure muscle mass, strength, and physical performance in community-dwelling older people: a systematic review. *Journal of the American Medical Directors Association*. 2013;14(3):170-8.
12. Bohannon RW, Peolsson A, Massy-Westropp N, Desrosiers J, Bear-Lehman J. Reference values for adult grip strength measured with a Jamar dynamometer: a descriptive meta-analysis. *Physiotherapy*. 2006;92(1):11-5.
13. McQuiddy VA, Scheerer CR, Lavalley R, McGrath T, Lin L. Normative values for grip and pinch strength for 6-to 19-year-olds. *Archives of physical medicine and rehabilitation*. 2015;96(9):1627-33.
14. Alidadi Y, Metanati M, Ataie-Jafari A. The validity of a bioelectrical impedance analyzer, Xiaomi MI scale 2, for measurement of body composition. 2019;2(2):36-8.
15. Kyle UG, Bosaeus I, De Lorenzo AD, Deurenberg P, Elia M, Gómez JM, et al. Bioelectrical impedance analysis—part I: review of principles and methods. *Clinical nutrition*. 2004;23(5):1226-43.
16. Sedgwick P. Spearman's rank correlation coefficient. *Bmj*. 2014;349.

17. Peplinkhuizen S, Eshuis G, Zijlstra WM, Timmer CY, Ploegstra MJ, Lelieveld OT, et al. Muscle strength is reduced in children with pulmonary arterial hypertension. *Pulmonary circulation*. 2023;13(2):e12246.
18. de Man Fd, Handoko M, Groepenhoff H, Van't Hul A, Abbink J, Koppers R, et al. Effects of exercise training in patients with idiopathic pulmonary arterial hypertension. *European Respiratory Journal*. 2009;34(3):669-75.
19. Laveneziana P, Albuquerque A, Aliverti A, Babb T, Barreiro E, Dres M, et al. ERS statement on respiratory muscle testing at rest and during exercise. *European Respiratory Journal*. 2019;53(6).
20. Gómez CA. Non-Invasive Monitoring of the Respiratory Muscles via Diffuse Optical Modalities: Boston University; 2024.
21. Calatayud J, Torres-Castro R, Vera-Urbe R, Olivares-Valenzuela Á, Guzmán-González B, Torres ME, et al. Neuromuscular and acute symptoms responses to progressive elastic resistance exercise in patients with chronic obstructive pulmonary disease: cross-sectional study. *Frontiers in Medicine*. 2022;9:934410.
22. Nagaya N, Nishikimi T, Okano Y, Uematsu M, Satoh T, Kyotani S, et al. Plasma brain natriuretic peptide levels increase in proportion to the extent of right ventricular dysfunction in pulmonary hypertension. *Journal of the American College of Cardiology*. 1998;31(1):202-8.
23. Lewis GD, Bossone E, Naeije R, Grünig E, Saggar R, Lancellotti P, et al. Pulmonary vascular hemodynamic response to exercise in cardiopulmonary diseases. *Circulation*. 2013;128(13):1470-9.
24. Mereles D, Ehlken N, Kreuscher S, Ghofrani S, Hoeper MM, Halank M, et al. Exercise and respiratory training improve exercise capacity and quality of life in patients with severe chronic pulmonary hypertension. *Circulation*. 2006;114(14):1482-9.
25. González-Saiz L, Fiuza-Luces C, Sanchis-Gomar F, Santos-Lozano A, Quezada-Loaiza CA, Flox-Camacho A, et al. Benefits of skeletal-muscle exercise training in pulmonary arterial hypertension: The WHOLEi+ 12 trial. *International journal of cardiology*. 2017;231:277-83.
26. 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. *Journal of physical therapy science*. 2015;27(5):1309-12.