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Original Article

Pulmonary arterial capacitance in children with pulmonary arterial hypertension and response to the treatment

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ABSTRACT

Objective: The pulmonary arterial capacitance index (PACi) has recently emerged as a dynamic marker of pulmonary vascular compliance. However, its clinical relevance in pediatric pulmonary arterial hypertension (PAH), and particularly its relationship with functional capacity and exercise tolerance in congenital heart disease (CHD), remains unclear. This study explored these associations and assessed changes following PAH-specific therapy.

Materials and Methods: Thirty-five patients with CHD-associated PAH with a mean pulmonary artery pressure (mPAP)≥20 mmHg receiving PAH-specific therapy and 35 age-, sex-, and anthropometry-matched CHD controls without PAH were evaluated. Demographic characteristics, hemodynamic parameters, PACi, functional class, brain natriuretic peptide (BNP) levels, and six-minute walk test (6MWT) distances were compared. Pre- and post-treatment hemodynamic and clinical parameters were also analyzed in the PAH group.

Results: Ventricular septal defect was the most common CHD in both groups. Children with PAH had significantly higher mPAP and pulmonary vascular resistance index (PVRi) and lower PACi than controls. PACi was inversely correlated with mPAP (r=-0.383, p=0.023) and PVRi (r=-0.812, p<0.01) but exhibited no significant association with BNP or 6MWT distance. No significant improvement in PACi or PVRi was observed after treatment.

Conclusion: PACi may serve as an early indicator of pulmonary vascular stiffness in pediatric PAH. Its limited association with functional and exercise-based assessments likely reflects early disease stages, age-related variability, and measurement constraints. Persistently low PACi and PVRi despite therapy underscore the progressive nature of pediatric PAH and highlight the need for larger, long-term prospective studies.

Keywords: congenital heart disease, exercise tolerance, functional capacity, pulmonary arterial capacitance, pulmonary arterial hypertension

INTRODUCTION

Pulmonary arterial hypertension (PAH) is a multifactorial and heterogeneous disorder defined by a mean pulmonary artery pressure (mPAP)≥20 mmHg, a pulmonary artery wedge pressure (PAWP)≤15 mmHg, and a pulmonary vascular resistance index (PVRi)≥3 Wood units·m² (WU·m²).[1-3] Early and accurate diagnosis, together with regular follow-up, is critical for guiding treatment decisions,

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assessing prognosis, and improving the quality of life of affected pediatric patients.[4]

PAH results in significant hemodynamic and histopathological changes in the pulmonary vasculature.[5] Parameters such as mPAP and PVRi have traditionally been used to evaluate these changes.[6] However, since these indices reflect only the static properties of pulmonary circulation, their prognostic value is limited.[7]

Consequently, attention has shifted toward dynamic parameters such as the pulmonary arterial capacitance index (PACi), defined as the ratio of stroke volume to pulmonary arterial pulse pressure and considered a measure of pulmonary vascular compliance.[8,9]

Adult PAH studies have associated reduced PACi with poor prognosis, and an inverse hyperbolic relationship between PACi and PVRi has been described. PVRi rises as PACi decreases, increasing right ventricular workload, hypertrophy, and ultimately right heart failure.[10,11] However, pediatric data regarding the relationship between PACi and exercise tolerance, functional capacity, and long-term outcomes remain limited.[8]

This study investigated the associations between PACi and PVRi, functional capacity, exercise tolerance, and serum brain natriuretic peptide (BNP) levels in children with PAH secondary to congenital heart disease (CHD) with left-to-right shunts. It also evaluated changes in these parameters before and after PAH-specific therapy.

A preliminary analysis of the research findings had previously been presented as a pediatric thesis (Yılmaz S. Pulmonary Arterial Capacitance in Children with Pulmonary Arterial Hypertension and Response to the Treatment. Gazi University, Faculty of Medicine, Department of Pediatric Health and Disease; 2014).[12]

MATERIALS AND METHODS

This retrospective study included pediatric patients diagnosed with CHD and PAH, who underwent diagnostic cardiac catheterization at the Gazi University, Faculty of Medicine, Department of Pediatric Cardiology, Türkiye. The study protocol was approved by the Gazi University Faculty of Medicine's Ethical Committee.

Study population

Data were collected from the hospital's electronic medical records and patient files. Patients were eligible if they were over three months of age, had a confirmed diagnosis of CHD, attended regular follow-up visits, and underwent diagnostic cardiac catheterization. Patients with persistent pulmonary hypertension of the newborn, idiopathic or heritable PAH, or neuromuscular disorders affecting functional assessment were excluded.

Two of the 40 patients with mPAP≥20 mmHg were excluded due to treatment non-compliance, two were lost to follow-up, and one had incomplete documentation. Thirty-five patients were included in the final analysis. A control group of 35 age-, sex-, and anthropometry-matched CHD patients without PAH, evaluated during the same period, was also included. These control patients underwent cardiac catheterization solely for shunt evaluation. Although they did not meet hemodynamic criteria for PAH, residual confounding related to underlying CHD physiology could not be completely excluded.

Treatment groups

The PAH patients were subdivided according to the treatment received:

- Monotherapy: Bosentan, iloprost, or sildenafil
- Combination therapy: Two or more PAH-specific drugs

Data collection

The following parameters were evaluated in the PAH group before and after therapy:

- Hemodynamic: mPAP, PVRi, and PACi
- Laboratory: serum BNP levels
- Clinical: six-minute walk test (6MWT) distance and New York Heart Association (NYHA) functional class
- Pediatric-specific: Ross classifications

Since the Ross classification does not routinely include direct measurements of peak oxygen consumption (%VO_2 max), this parameter was excluded from the analysis.

Pre-treatment values were obtained from the first catheterization at admission, while post-treatment values were obtained from follow-up catheterizations. Children younger than six years or physically unable to perform the 6MWT were excluded from exercise capacity evaluation. Fourteen children were eventually excluded from the 6MWT assessment because they were under 6 years of age.

Hemodynamic calculations

Hemodynamic data recorded during catheterization were used to calculate stroke volume and PVRi:[13]

Stroke volume (SV) (mL)=Cardiac output (CO) / Heart rate (HR)

The following formula was used to calculate the PACi, representing the ratio of volume change to pressure change in the pulmonary artery:[9]

PACi (mL/mmHg/m²)=Stroke volume (SV) / Pulmonary artery pulse pressure (systolic – diastolic PA pressure) / Body surface area (m²)

Statistical analysis

Data were analyzed using Statistical Package for the Social Sciences software (SPSS, IBM Corp., Armonk, NY, USA). Descriptive statistics were expressed as mean ± standard deviation (SD), median (minimum–maximum), frequency, and percentage values. Due to wide value ranges in some variables, non-parametric methods were predominantly used.

The normality of data distribution was assessed using both visual methods (histograms and probability plots) and analytical tests (Kolmogorov-Smirnov and Shapiro-Wilk). The Mann-Whitney U test was applied for non-normally distributed continuous variables in intergroup comparisons, while the chi-square test was used for categorical variables.

Pre- and post-treatment comparisons of continuous variables were performed using the Wilcoxon signed-rank test. Functional class comparisons using the modified Ross and NYHA classifications were evaluated with the McNemar-Bowker test and the Wilcoxon test.

Spearman's correlation analysis was conducted to assess relationships among PACi, mPAP, PVRi, BNP levels, and 6MWT distance.

A p-value of<0.05 was considered statistically significant.

RESULTS

Thirty-five patients were included in both the PAH and control groups. The control group consisted of 16 girls (45.7%) and 19 boys (54.3%), and the PAH group of 13 girls (37.1%) and 22 boys (62.9%). The most common congenital heart defect in both groups was a ventricular septal defect (Table 1).

Table 1. Distributions of Congenital Heart Defects in Patients with PAH and the Control Group

	Patient Group n=35 (%)	Control Group n=35 (%)
ASD	2 (5.7%)	10 (28.6%)
VSD	19 (54.2%)	16 (45.8%)
PDA	1 (2.9%)	6 (17.1%)
ASD+VSD	3 (8.5%)	-
ASD+VSD+PDA	3 (8.5%)	-
AVSD	1 (2.9%)	1(2.8%)
PFO	-	1(2.8%)
VSD+PFO+BAV	-	1(2.8%)
PA+ASD	1 (2.9%)	-
AP Window	1 (2.9%)	-
TAPVR+ASD	1 (2.9%)	-
AVSD+Pulmonary artery anomalies	1 (2.9%)	-
TGA+ASD+VSD	1 (2.9%)	-
TGA+Pulmonary artery banding	1 (2.9%)	-

Abbreviations: ASD: Atrial Septal Defect, VSD: Ventricular Septal Defect, PDA: Patent Ductus Arteriosus, AVSD: Atrioventricular Septal Defect, PFO: Patent Foramen Ovale, BAV: Bicuspid Aortic Valve, PA: Pulmonary Atresia, AP Window: Aortopulmonary Window, TAPVR: Total Anomalous Pulmonary Venous Return, TGA: Transposition of the Great Arteries.

Pre-treatment age, weight, and body surface area were significantly lower in the PAH group than in the controls (p=0.032, p=0.033, and p=0.033, respectively). All three parameters increased significantly after treatment. In terms of hemodynamics, the PAH group exhibited significantly higher systolic, diastolic, and mean pulmonary artery pressure (mPAP), and pulmonary vascular resistance index (PVRi), and lower pulmonary arterial capacitance index (PACi) than controls (p<0.05). The post-treatment cardiac index decreased significantly within the PAH group (p=0.006), while PACi remained lower than in the controls (Table 2).

Table 2. Patients' Pre- and Post-Treatment Demographic Characteristics and Hemodynamic Parameters				
		Patient Group	**Control Group**	**p1/p2**
Age (months)	Pre-treatment	48 (6-212)	132 (3-204)	0.032
	Post-treatment	85 (17-216)	132 (3-204)	0.541
		p^3=0.001		
Height (cm)	Pre-treatment	100 (63-177)	132 (67-139)	0.060
	Post-treatment	113 (75-177)	132 (67-139)	0.492
		p^3=0.001		
Weight (kg)	Pre-treatment	15 (5-75)	28 (5-63)	0.033
	Post-treatment	19.5 (7-76)	28 (5-63)	0.417
		p^3=0.001		
Body surface area (m^2)	Pre-treatment	0.62 (0.27-1.73)	1.01 (0.28-1.69)	0.033
	Post-treatment	0.77 (0.37-1.73)	1.01 (0.28-1.69)	0.414
		p^3=0.001		
Cardiac index (L/m^2)	Pre-treatment	7.24 (0.93-62.96)	4.94 (2.54-28.33)	0.285
	Post-treatment	5.56 (0.93-26.67)	4.94 (2.54-28.33)	0.643
		p^3=0.006		
Systolic PAP (mmHg)	Pre-treatment	98 (52-125)	25 (18-34)	0.001
	Post-treatment	102 (44-130)	25 (18-34)	0.001
		p^3=0.748		
Diastolic PAP (mmHg)	Pre-treatment	46 (11-80)	9 (5-14)	0.001
	Post-treatment	47 (9-90)	9 (5-14)	0.001
		p^3=0.738		
Mean PAP (mmHg)	Pre-treatment	70 (30-95)	16 (12-21)	0.001
	Post-treatment	71 (22-107)	16 (12-21)	0.001
		p^3=0.707		
PVRi (WU x m^2)	Pre-treatment	6.71 (0.66-65.61)	1.4 (0.15-2.61)	0.001
	Post-treatment	8.6 (0.82-58.02)	1.4 (0.15-2.61)	0.001
		p^3=0.028		
PACi ($mL/mmHg/m^2$)	Pre-treatment	1.5 (0.192-10.678)	2.91 (1.97-366.06)	0.001
	Post-treatment	1.1 (0.192-6.213)	2.91 (1.97-366.06)	0.001
		p^3=0.077		
Qp ($L/min/m^2$)	Pre-treatment	4.8 (1.61-17)	4.9 (3.2-17)	0.259
	Post-treatment	4.8 (1.61-12)	4.9 (3.2-17)	0.109
		p^3=0.193		
Qs ($L/min/m^2$)	Pre-treatment	3.5 (1.82-23.77)	3.8 (2.9-9.4)	0.251
	Post-treatment	3.5 (1.1-5.65)	3.8 (2.9-9.4)	0.078
		p^3=0.669		

p<0.05 Mann-Whitney U, p^1: Pre-treatment PAH patients-control group, p^2: Post-treatment PAH patients-control group, p^3: Pre-treatment- post-treatment PAH PAP: Pulmonary artery pressure, PVRi: Pulmonary vascular resistance index, PACi: Pulmonary artery capacitance index, Qp: Pulmonary blood flow, Qs: Systemic blood flow

Table 3. A comparison of BNP levels and six-minute walk test results between the patient and control groups

		Patient group median (min-max)	Control group	p1/p2
BNP levels	Pre-treatment	650 (60-3000)	35 (10-65)	0.001
	Post-treatment	498 (60-1288)	35 (10-65)	0.001
		p^3=0.328		
6-MWT values	Pre-treatment	420 (153-560)	540 (510-585)	0.001
	Post-treatment	450 (300-564)	540 (510-585)	0.001
		p^3=0.123		

p<0.05 Mann-Whitney U, p^1=Comparison between the pre-treatment group and the control group; p^2=Comparison between the post-treatment group and the control group; p^3=Comparison between pre-treatment and post-treatment values within the patient group.

BNP levels and Six-Minute Walk Test (6-MWT)

Due to the retrospective nature of the study, pre-treatment BNP levels were only available for 14 patients, and post-treatment levels for 21. BNP analyses were conducted using available-case data, and patients with missing BNP values were excluded from the relevant comparisons.

The BNP levels of the PAH group, both before and after PAH-specific therapy, differed significantly from those of the control group (p<0.05 for both; Table 3). Similarly, significant differences were observed between the PAH and control groups' 6MWT results at both time points (p< 0.05 for both).

Fourteen children were younger than 6 years and, therefore, ineligible to perform the 6MWT, which restricted exercise capacity analyses to the remaining patients.

However, no statistically significant difference was observed between pre- and post-treatment BNP levels or 6MWT distances within the PAH group (p>0.05) (Table 3).

Functional capacity

Functional capacity, assessed using the modified Ross and NYHA classifications, showed no significant differences between pre- and post-treatment evaluations (p=0.617 and p=0.123, respectively). Table 4 summarizes the distribution of functional classes. The majority of patients in the PAH group were in Ross Class II before (71.4%) and after treatment (71.4%), with only minor shifts between Class I and Class III. Similarly, NYHA classifications showed that the majority of patients remained in Class II at both time points (74.3% pre-treatment and 74.3% post-treatment). Only small proportions of patients were classified as Class I or III in either system, and no statistically significant changes were observed after therapy (p>0.05 for both the Ross and NYHA systems).

Table 4. Number of patients in the Ross and NYHA Classifications in the patient and control groups

		Pre-treatment	Post-treatment	p
Ross Classification	Class I	7	8	
	Class II	25	25	
	Class III	3	2	
	Ross	2 (1-3)	2 (1-3)	0.617
NYHA Classification	Class I	4	7	
	Class II	26	26	
	Class III	5	2	
	NYHA	2 (1-3)	2 (1-3)	0.109

p<0.05 Mann-Whitney U, p^1=Comparison of the Ross and NYHA classifications between the pre-treatment patient group and the control group, p^2=Comparison of the Ross and NYHA classifications between the post-treatment patient group and the control group, p^3=Comparison of the Ross and NYHA classifications between the patient group

Correlation analysis

The relationships between PACi, PVRi, and mPAP values and serum BNP levels and 6MWT results in the patient group were assessed using Spearman correlation analysis (Table 5).

Table 5. Correlations between PACi, mPAP, and PVRi values and BNP levels and 6-MWT distance in the patient groups

	mPAP	PACi	PVRi	BNP	6-MWT
PACi	r=-0.383* p=0.023	r=1.000 p=-	r=-0.812** p<0.01	r=0.4 p=0.072	r=0.139 p=0.701
PVRi	r=0.669** p<0.01	r=-0.812** p<0.01	r=1.000 p=-	r=-0.326 p=0.149	r=-0.588 p=0.074
mPAP	r=1.000 p=-	r=-0.383* p=0.023	r=0.669** p<0.01	r=-0.129 p=0.577	r=- 0.116 p=0.751
BNP	r=- 0.129 p=0.577	r=0.400 p=0.072	r=-0.326 p=0.149	r=1.000 p=-	r=-0.101 p=0.848
6-MWT	r=-0.116 p=0.751	r=0.139 p=0.701	r=-0.588 p=0.074	r=-0.101 p=0.848	r=1.000 p=-

*Significant at the 0.05 level (2-tailed)
**Significant at the 0.01 level (2-tailed)
mPAP: mean Pulmonary artery pressure, PACi: Pulmonary artery capacitance index, PVRi: Pulmonary vascular resistance index, BNP: Brain Natriuretic Peptide, 6-MWT: six-minute walk test.

Analysis revealed a statistically significant negative correlation between PACi and mean pulmonary artery pressure (mPAP) at the 0.05 significance level (r=–0.383, p=0.023), and between PACi and PVRi at the 0.01 level (r=–0.812, p<0.01). No significant correlation was observed between PACi and either BNP levels or 6MWT results.

Additionally, PVRi was positively correlated with mPAP at the 0.01 significance level (r=0.669, p<0.01) and negatively correlated with PACi at the 0.05 significance level. However, no significant correlation was observed between PVRi and either BNP or 6MWT values.

DISCUSSION

Pulmonary arterial hypertension remains a major cause of morbidity and mortality in both adults and children, particularly in those with CHD. As reported in previous pediatric cohorts, a ventricular septal defect was the most common lesion in our study population.[14]

The management of pediatric PAH requires accurate and dynamic prognostic markers. Traditional parameters such as mPAP and PVRi provide essential information but predominantly reflect static vascular properties.[15] In the present cohort, PVRi increased significantly after treatment despite stable functional capacity, suggesting that conventional markers may not fully capture early hemodynamic deterioration.

Pulmonary arterial capacitance index (PACi) has emerged as a dynamic indicator of pulmonary vascular compliance.[7-9] Consistent with previous studies, children with PAH in our cohort demonstrated significantly reduced PACi values compared with the controls, and PACi exhibited inverse correlations with mPAP and PVRi.[16] Recent pediatric data indicate that impaired pulmonary arterial compliance independently predicts short-term clinical deterioration, underscoring the prognostic relevance of compliance-based indices in childhood PAH.[17] Importantly, PACi did not improve after therapy. Taken together, these findings indicate that despite treatment, low PACi persisted, suggesting ongoing vascular remodeling and loss of arterial compliance.[18,19] The lack of improvement in PACi and PVRi after therapy may be attributable to several factors. Treatment escalation or optimization was not uniform across all patients, and some children remained on monotherapy due to age, tolerability, or clinical stability. Additionally, the follow-up intervals may have been insufficient to capture structural vascular changes, since pulmonary vascular remodeling often progresses slowly and may not be detectable over short-term catheterization periods. Some patients may also have had more advanced vascular disease at baseline, limiting the potential for measurable reversibility.

Biomarkers such as BNP and NT-proBNP are widely used for monitoring PAH and have been linked to hemodynamic status and exercise capacity.[20-22] In this study, no significant change in BNP values was observed after treatment, likely due to the limited sample size and incomplete data.

Previous retrospective evaluations of complications and cardiac risk factors have similarly been limited by small sample sizes, supporting the need for larger, prospective studies.[23] Nevertheless, a negative correlation between BNP levels and 6MWT distance is suggestive of its role as an indicator of functional limitation.

Although adult studies have demonstrated that lower PVRi and higher PACi are associated with longer 6MWT distances, our analysis showed no correlation between PACi and 6MWT or BNP levels.[24] This discrepancy may be attributed to age-related differences in exercise performance, limited patient numbers, and non-synchronous data collection. Larger studies with standardized assessments are now needed to clarify these relationships. Several factors may further explain the lack of association between PACi and BNP or 6MWT in this cohort. The limited size and incomplete BNP dataset substantially reduced statistical power, limiting our ability to detect moderate correlations. Additionally, BNP measurements, cardiac catheterization data, and 6MWT results were not collected synchronously, potentially masking short-term physiological changes in pulmonary vascular compliance. Exercise performance in children also exhibits wide age-related variability. Differences in stride length, motivation, cooperation, and developmental stage, particularly among younger children, can introduce significant noise into functional assessments. These methodological and age-related factors likely contributed to the absence of measurable correlations in this study.

The Ross and NYHA classifications were used to measure functional capacity in this study. The majority of patients were in FC II, and no significant functional improvement was observed during follow-up. The limited change in functional class may reflect several factors specific to pediatric PAH.[25,26] In contrast to adults, as noted in the REVEAL registry, children frequently exhibit variations in functional scores despite alterations in hemodynamic or biomarker parameters. Age-related differences in activity perception, developmental variability in exercise tolerance, and the inherently subjective nature of these scoring systems may all contribute to this discrepancy. In younger children, caregiver-reported assessments may obscure subtle changes, whereas older children may adapt to chronic symptoms and underreport limitations. Additionally, the relatively short follow-up period and small sample size in the present study may have reduced the likelihood of detecting clinically meaningful changes in functional class. Prospective, multicenter studies are now needed to validate pediatric functional scoring tools and to better define their role in monitoring disease progression and treatment response.

Study limitations

This study has several limitations. First, its retrospective design restricted access to complete clinical, laboratory, and functional data, particularly concerning BNP levels. The incomplete BNP dataset reduced the strength of correlation and longitudinal analyses and limited the interpretability of biomarker-hemodynamic relationships. Second, the relatively small sample size (n=35) substantially reduced statistical power and limited our ability to perform subgroup analyses, which should be considered when interpreting the findings. These factors may have affected the robustness and generalizability of the results. Additionally, since the control group consisted of CHD patients undergoing catheterization solely for shunt evaluation, residual confounding related to underlying cardiac physiology cannot be completely excluded. Further prospective, multicenter studies with larger and more homogeneous cohorts and standardized data collection are now needed to validate the study.

CONCLUSION

Pulmonary arterial hypertension in children remains a progressive condition despite targeted therapies, reflecting ongoing vascular remodeling and limited treatment options. Early identification of hemodynamic changes, particularly using parameters such as PACi, may enhance clinical assessment and guide management strategies.

Future prospective multicenter studies with larger cohorts are essential to clarify disease mechanisms and support the development of novel, targeted therapies for pediatric PAH.

Ethical approval

This study has been approved by the Gazi University Faculty of Medicine's Ethical Committee (approval date 2013, decision no. 2013-111).

Author contribution

The authors declare contribution to the paper as follows: Study conception and design: SO; data collection: SO, SK; analysis and interpretation of results: SO; draft manuscript preparation: SO, SK. All authors reviewed the results and approved the final version of the article.

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Conflict of interest

The authors declare that there is no conflict of interest.

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