

INTRODUCTION

Pulmonary arterial hypertension (PAH) is a progressive disease that has high mortality and morbidity. Although PAH-targeted therapies, outcome of PAH is still poor, especially in children. Despite the 6 minute walk test (6MWT) used in the follow-up of patients with PAH has been shown to have prognostic value in adults, data is limited in children because of difficulties in performing test. As well as there are studies with conflicting results regarding the prognostic value of 6MWT, there are only a few studies in the literature that have examined the predictive values of saturation and heart rate during the test in children. This study aims to determine the prognostic values of heart rate (HR) and transcutaneous oxygen saturation (Sat) which were measured before, at the end of and 5 minutes after 6MWT in children who are undergoing PAH specific treatment.

MATERIALS & METHODS

This is a prospective study of 29 children who were aged between 7 and 17 years, diagnosed with PAH and underwent 6MWT between 2006 and 2015 in Department of Pediatric Cardiology, Gazi University. PAH is defined as; right cardiac ventricle catheterization determined as mean pulmonary artery pressure (mPAP) ≥ 25 mmHg, pulmonary capillary wedge pressure (PCWP) ≤ 15 mmHg, PVR index (PVRi) ≥ 3 $wu \cdot m^2$. The 6MWT test is conducted according to the guidelines reported by the American Thoracic Society.

Transcutaneous oxygen saturation and heart rate were measured before 6MWT, at the end of 6MWT and 5 minutes after the completion of 6MWT using a handheld pulse oxymeter placed on the index finger of the patient's right hand. (Sat₀, Sat₁, Sat₂; HR₀, HR₁, HR₂ respectively). HR increase was defined as HR₁ minus HR₀ and decrease in oxygen saturation was defined as Sat₀ minus Sat₁.

RESULTS

15 of 29 patients (51,9%) were female. The mean age of the children was 129 \pm 45 months and the mean follow-up time was 58 \pm 40 months. The most common etiology of PAH was ventricular septal defect (VSD) (38%). Primary PAH was diagnosed in 2 patients. The mean proBNP concentrations did not change significantly before and after the administration of PAH specific treatment (946.4 \pm 1754.8 pg/ml vs 917.6 \pm 1920 pg/ml p=0.11). However, the mean 6MWT distance increased significantly after the administration of PAH specific treatment (400.2 \pm 107.8 m vs 436.2 \pm 119.2 m, p=0.012). The 6MWT distance was significantly longer and HR₁ was significantly higher in survivors (453.3 \pm 96.5 m vs 250 \pm 135.2 m, p=0.025 and 122.8 \pm 18.4 /min vs 94.3 \pm 19.1 /min, p=0.034). HR₁ increased significantly but Sat₁ and Sat₂ decreased significantly after PAH specific treatment (p=0.017, p=0.03 and p=0.017 respectively) (Table 1).

	Before treatment	After treatment	p
	mean \pm s.d.	mean \pm s.d.	
proBNP (pg/ml)	946.4 \pm 1754.8	917.6 \pm 1920	0.11
6MWT (m)	400.2 \pm 107.8	436.2 \pm 119.2	0.012
Mean PAP (mmHg)	58.7 \pm 25.9	73 \pm 29.7	
Saturation (%)			
0	87.3 \pm 12.3	86.6 \pm 9.1	0.47
1	76.2 \pm 21.6	65 \pm 26.7	0.003
2	81.8 \pm 16.6	71.7 \pm 21.3	0.017
HR (/min)			
0	96.5 \pm 15.9	95.2 \pm 13.7	0.81
1	118.9 \pm 20.4	132.3 \pm 22.7	0.017
2	106.1 \pm 17.4	114.5 \pm 21.4	0.26
FC (%)			
I		13.8	
II	20.7	55.2	
III	62.1	24.1	
IV	17.2	6.9	
Monotherapy (n)	11	9	
Combined therapy (n)	18	17	

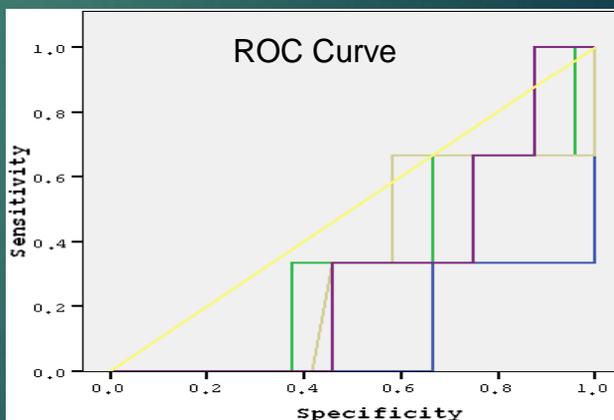
While there was no significant correlation between World Health Organization functional classification (WHO-FC) and Sat₁ before treatment, WHO-FC and Sat₁ correlated negatively after treatment (r=-0.435, p=0.021). Although WHO-FC and proBNP did not correlate before treatment, they correlated positively after treatment (p=0.0001) (Table 2).

		WHO Functional Class	
		Before Treatment	After Treatment
Saturation 1	r	-0.35	-0.43
	p	0.06	0.02
proBNP	r	0.3	0.6
	p	0.1	0.0001

The correlation between 6MWT and HR, Sat before/after treatment is shown in Table 3.

		6MWT	
		Before Treatment	After Treatment
Heart Rate	0	r	-0.37
		p	0.04
	1	r	-0.13
		p	0.49
	2	r	-0.43
		p	0.02
Saturation	0	r	0.36
		p	0.05
	1	r	0.47
		p	0.009
	2	r	0.67
		p	0.0001

There was no association between survival and heart rate increase whereas survival was positively associated with the decrease in oxygen saturation. If the decrease in oxygen saturation was 14.6%, sensitivity increased to 100% and specificity was 58.3% (Graphic 1).



CONCLUSION

PAH is a chronic disease which continues to progress despite the administration of specific therapy. The measurement of oxygen saturation with pulse oximeter during 6MWT might have prognostic importance in the follow up of children undergoing PAH specific treatment.