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The Etiology and Clinical Characteristics of Patients with Pulmonary Hypertension without Congenital Heart Disease

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Pulmonary hypertension (PH) is a rare progressive disorder in children that is associated with significant morbidity and mortality. Most common causes of PH in children include; congenital heart diseases, idiopathic pulmonary arterial hypertension, and rarely; connective tissue disorders and thromboembolism. We aimed, in our study, to evaluate the etiology and clinical features of study population that are not associated with congenital heart diseases other than the small atrial septal defects not causing serious PH.

METHODS AND RESULTS:

From 2011 to 2016, 180 pediatric PH patients were evaluated for the clinical features and etiologic distrubition of PH. 37 patients with idiopathic pulmonary hypertension (IPAH) and PAH associated with other diseases were enrolled in our study. The baseline characteristics were detailed in *Table 1*. 20 patients with IPAH, and 17 patients with PAH associated with other diseases were studied. Median age for IPAH was lower than PAH associated with other disease (4,7 to 7,2 years). Patients with PAH associated with other diseases included chronic renal failure (n=2), pulmonary disease (n=7), portopulmonary hypertension (n=1) veno-oclusive disease (n=1) and systemic juvenile idiopathic arthritis (n=1). Syndromes associated with PAH were Down syndromes (n=2), Allagille syndrome (n=1), Dursun syndrome (congenital neutropenia, n=1) and Cleidocranial Dysostosis (n=1). Six patients associated with pulmonary disease, three patients had interstitial lung disease, one patient had bronchiolitis obliterans, one patient had bronchopulmonary dysplasia and other one had obstructive sleep apnea syndrome. In IPAH patients, two patients had hematologic comorbidities (thalassemia carrier and hypofibrinogenemia).

DISCUSSION:

Pulmonary hypertension is a progressive disease with a poor diagnosis. Due to lack of specific symptoms and signs especially in the early stage of the disease, the diagnosis is usually delayed. A systemic approach considering the coexistent disorders is required for the diagnosis. Hence, systemic classification is essential in determining the high-risk groups who should be screened.

	IPAH (n=20)	PAH associated with other diseases (n=17)
Age at diagnosis (years)	4,7 (0,5-14)	7,2 (0,1-17,7)
Gender; female	13 (65%)	8 (47%)
Comorbities, (n)		
Secundum ASD	9	3
Syndrome	-	5
Obesite	-	1
Pulmonary	-	6
Hematologic	2	1
Gastrointestinal	-	2
Renal	-	2
Hemodynamics at diagnosis		
Catheterization	n=18	n=12
PVRi (WU.m2)	15,4	13,8
PVR/SVR	61,1	46,6
Mean PAP (mmHg)	60±16,6	42±16
Echocardiographic[1]	n=2	n=5
PAP	87,5±3,5	51±19,8
Pulmonary Therapy, n		
Anti congestive therapy	-	5
Monotherapy*	10	4
Combination	6°, 2ˣ	6°, 1ˣ
Triple Therapy	2ᵝ	1ᵝ

TABLE 1 IPAH= Idiopathic pulmonary artery hypertension, PAH= pulmonary artery hypertension, ASD= atrial septal defect, PVRi= Pulmonary Vascular Resistance index, PVR/SVR= ratio of pulmonary vascular resistance to systemic vascular resistance, PAP= pulmonary arterial pressure, WU= Woods units, [1] Measurement of the peak tricuspid regurgitant velocity, *ERAs (Endothelin receptors antagonist); ° ERAs and PDE-5 inh(phosphodiestere-5 inhibitors); ˣ ERAs and PGI2 (Prostacyclin analogue); ᵝ ERAs, PDE-5 and PGI2