Pulmonary arterial hypertension in Spanish pediatric registry age: clinical characterization, management and survival

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Background: Pulmonary arterial hypertension (PAH) includes idiopathic PAH and congenital heart disease (CHD) related PAH. A wide variety of CHD can develop PAH, but their clinical characteristics define four large groups: Eisenmenger, PAH associated with non-restrictive shunt, severe PAH associated with restrictive shunt and postoperative PAH. Our aim was to study the clinical and prognostic differences between these groups and idiopathic PAH.

Methods: The REHIPED registry is a Spanish, multicenter, observational and prospective registry on pulmonary hypertension in the pediatric population (<18 years of age) that began in 2008. 183 patients were analyzed: baseline characteristics, functional class, right catheterization data, treatment and survival were compared

Results: 183 patients were analyzed, characteristics are detailed in the table. In patients with idiopathic PAH, treatment with prostanoids was more frequently used as first line therapy and during follow up. The average follow-up time was 9 years. There was not a statistically significant difference in survival among the 4 clinical groups of CHD related PAH. There was a borderline significant difference (logrank p 0.05) in survival between the group of CHD related PAH and idiopathic PAH.

Conclusion: Idiopathic PAH patients have worse outcome than CHD related PAH patients although they have less comorbidities, less severe hemodynamics and are treated more aggressively.

	Total	Eisenmenger	Non restrictive shunts	Restrictive shunts	Postoperative PAH	Idiopathic PAH	p
N, n (%)	146	31 (16)	38 (20)	8 (4)	67 (35)	47 (25)	
Sex (female), n (%)	81 (56)	17 (55)	26 (68)	4 (50)	34 (51)	23 (49)	0.714
Age, mean ± SD	4.8±4.7	8.9±4.6	3.2±4.3	2.4±4.1	4.5±4.7	4.5±3.6	<0.001
WHO Functional class III–IV, n (%)	64 (43,8)	15 (48)	15 (39)	2 (25)	34 (51)	17 (36)	0.389
Chromosomopathy, n (%)	62 (32.5)	15 (48)	13 (34)	2 (25)	30 (44.8)	2 (4.3)	<0.001
Mean pulmonary arterial pressure (mmHg), mean ± SD	45.5±17.6	62.1±17.3	39.8±12.7	40±18	42±18	47.3±16.3	<0.001
Pulmonary vascular resistance index (uW·m^2), mean ± SD	9.9±8.4	15.4±9.7	5.2±3	8.5±6	8.9±8.9	12±8.3	<0.001
Initial therapy							0.005
Oral monotherapy, n (%)	146 (83)	29 (97)	30 (91)	4 (57)	52 (88)	31 (67)	
Monotherapy with prostanoids, n (%)	7 (5)	0	1 (3)	2 (29)	0	5 (11)	
Oral combination therapy, n (%)	14 (10,2)	1 (3,3)	2 (6)	1 (14)	6 (10)	6 (13)	
Combination therapy with prostanoids, n (%)	5 (4)	0	0	0	1 (2)	4 (9)	
Prostanoids during follow up, n (%)	52 (27)	4 (13)	5 (13)	3 (38)	13 (19)	27 (57)	<0.001