Pulmonary arterial hypertension associated with congenital heart disease in children and adults: knowing the present to imagine the future

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Introduction: Pulmonary arterial hypertension (PAH) is a common comorbidity in congenital heart disease (CHD) and significantly affects prognosis. Children born in recent years with CHD have benefited from advances in pediatric cardiac surgery that have modified their evolution compared to adults with the same pathologies.

Purpose: Our objective was to define and compare the characteristics of the adult and pediatric population with PAH associated with CHD.

Methods: REHAP and REHIPED are Spanish, voluntary, multicenter registries that include patients with PAH associated with CHD. REHAP started in 2007 and includes patients over 18 years of age. Patients diagnosed with PAH after this time were prospectively included, until 2020. Patients who were followed in the participating centres and diagnosed after January 1998 were retrospectively included in the registry. REHIPED started in 2009 and collects patients between 2 months and 18 years of age. Patients diagnosed with PAH after this time were prospectively included, until 2020. Patients who were followed and diagnosed after January 1998 were retrospectively included. The baseline characteristics, hemodynamic profile, treatment, and 10-year survival of the REHAP patients were compared to those included in the REHIPED. In addition, the change in the distribution of etiologies of REHAP over time was analyzed.

Results: 664 patients were included from REHAP (mean age 32±20 years) and 153 from REHIPED (mean age 5±5 years). Among adults, there was a significantly higher proportion of women (67% versus 56%, p 0.004), patients with Eisenmenger syndrome and simple heart disease. The frequency of the different clinical groups has varied over the years, becoming progressively similar to the distribution observed in REHIPED. Adults presented greater hemodynamic severity, with higher mean pulmonary arterial pressure and greater pulmonary vascular resistance index. There were no differences in initial therapy or in the percentage of patients who received prostanoids at follow-up. Survival at 10 years was better in the group of children (99% at 1 year, 95% at 5 and 80% at 10) than in the adult group (94% at 1 year, 83% at 5 and 71% at 10) with a HR of 1.93 [95% CI 1.2–3] for REHAP compared to REHIPED.

Conclusions: Patients with PAH associated with CHD have different clinical and prognostic characteristics in childhood and adulthood. Knowing the characteristic of the pediatric age can help predict how the future patients with PAH associated with CHD will be and how we can plan their care.

	REHAP	REHIPED	p
N	664	153	
Sex (female)	447 (67)	84 (56)	**0,004**
Age (years)	32 ± 20	5 ± 5	**<0,001**
Clinical group			**<0,001**
Eisenmenger, n(%)	371 (56)	32 (21)	
Large left to right shunt, n(%)	68 (10)	42 (28)	
Small shunt, n(%)	62 (9)	8 (5)	
Postoperative, n(%)	159 (24)	68 (45)	
Type of defect			**<0,001**
Simple, n(%)	415 (63)	50 (39)	
Complex, n(%)	167 (25)	38 (30)	
Combined, n(%)	82 (12)	40 (31)	
Down Syndrome	132 (20)	47 (31)	**0,004**
NYHA III-IV, n(%)	331 (50)	60 (45)	0,289
Right atrial pressure (mmHg)	9 ± 5	10 ± 5	0,4389
Mean pulmonary artery pressure (mmHg)	60 ± 19	45 ± 17	**<0,001**
Pulmonary vascular resistance index (Wood Units m ²)	18 ± 12	9 ± 8	**<0,001**
First line therapy			0,108
Oral monotherapy	473 (86)	121 (89)	
Oral combination therapy	30 (5)	11 (8)	
Combination therapy with prostanoids	11 (2)	1 (0,7)	
Prostanoids at any time during follow up	134 (20)	26 (17)	0,370

All data are expressed as mean ± SD or n (%). NYHA: New York Heart Association

Table 1

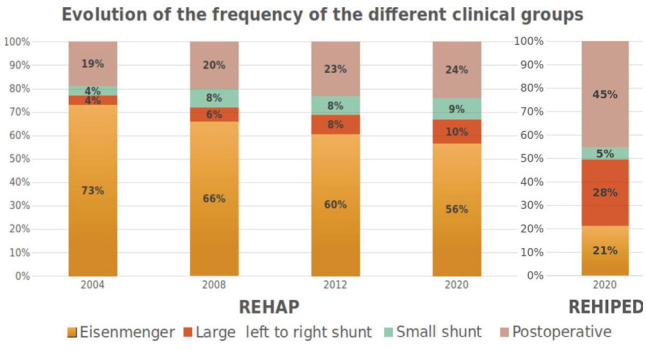

Figure 1