Pulmonary hypertension and congenital heart disease: medical treatment and risk factors for survival

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On behalf of REHAP investigators

Funding Acknowledgement: Type of funding source: Private grant(s) and/or Sponsorship. Main funding source(s): Ferrer, Janssen

Background: Pulmonary arterial hypertension (PAH) is a common comorbidity in congenital heart disease (CHD) and significantly affects prognosis. There are four large clinical groups of CHD-related PAH: Eisenmenger syndrome, PAH associated with non-restrictive shunt, PAH associated with restrictive shunt and postoperative PAH. Our purpose was to study the clinical and prognostic differences among them.

Methods: The REHAP is a Spanish multicentre voluntary registry of patients over 14 years of age, which includes patients with CHD and PAH, starting in 2007. 664 patients were analyzed: Baseline characteristics, functional class, right catheterization data, treatment and survival were compared.

Results: 664 patients were analyzed: characteristics are detailed in the table. Patients with Eisenmenger were more frequently treated with oral monotherapy as a first line therapy and received less frequently prostanoids during the follow-up. Patients with Eisenmenger had significantly better prognosis, with the best long-term survival of the 4 groups. In a cox regression model, postoperative PAH has a 1.7 hazard ratio (HR) (reference group: Eisenmenger) after adjustment for age (HR 1.02 p 0,001), functional class (HR NYHA III-IV 2.3 p<0,001), sex (p 0.8) and pulmonary vascular resistance (p 0.7).

Conclusion: The clinical classification of PAH associated with CHD defines both the baseline characteristics and the prognosis of patients. Outcome relates closely to functional class and type of PAH–CHD. Eisenmenger group, which has the most severe hemodynamics, is the one with the best prognosis despite a less aggressive treatment.

	Eisenmenger	Large left to right shunt	Small shunt	Postoperative	All	p
N, n%	371 (56)	68 (10)	62 (9)	159 (24)	660	
Age (years), mean ± SD	34±16	46±17	46±19	40±17	38±17	<0.001
Sex (female), n (%)	236 (64)	52 (76)	45 (73)	112 (70)	445 (67)	0.094
NYHA Functional class III–IV, n (%)	191 (51)	36 (53)	31 (50)	71 (45)	329 (50)	0.499
Right atrial pressure (mmHg), mean ± SD	8±5	9±4	9±5	10±6	9±5	0.001
Pulmonary vascular resistance (Wood Units), mean ± SD	14±7	10±8	9±5	10±7	11±7	<0.001
First line therapy						0.001
Oral monotherapy, n (%)	277 (90)	38 (78)	44 (83)	112 (81)	471 (86)	
Oral combination therapy, n (%)	8 (3)	7 (14)	7 (13)	8 (6)	30 (5)	
Combination therapy with prostanoids, n (%)	5 (2)	1 (2)	2 (4)	3 (2)	11 (2)	
Prostanoids at any time during follow up, n (%)	63 (17)	10 (15)	16 (25)	45 (28)	134 (20)	0.01

NYHA: New York Heart Association; 6MWD: six-minute walking distance.