Idiopathic pulmonary hypertension in Spanish pediatric registry: clinical characterization, management, and risk factors for survival

J. Playan Escribano[1], T. Segura De La Cal[2], P. Escribano Subias[2], A. Mendoza Soto[2], C. Labrandero[3], I. Guillen Rodriguez[4], L.M. Caicedo Cuenca[5], A. Sabate Rotes[6], S. Villagra Albert[7], M. Lopez Ramon[8], L. Conejo Munoz[9], M.A. Izquierdo Riezu[10], G. Inigo Martin[11], S. Escriba Bori[12], M.J. Del Cerro Marin[13]

[1]Hospital Clinico San Carlos, Madrid, Spain; [2]University Hospital 12 de Octubre, Madrid, Spain; [3]University Hospital La Paz, Madrid, Spain; [4]University Hospital of Virgen del Rocio, Seville, Spain; [5]Shaio Clinic Foundation, Bogota, Colombia; [6]University Hospital Vall d'Hebron, Barcelona, Spain; [7]Congenital Heart Diseases Unit (UCC), Madrid, Spain; [8]University Hospital Miguel Servet, Zaragoza, Spain; [9]Regional University Hospital of Malaga, Malaga, Spain; [10]Donostia University Hospital, San Sebastian, Spain; [11]Hospital Virgen de la Salud, Toledo, Spain; [12]University Hospital Son Espases, Palma de Mallorca, Spain; [13]University Hospital Ramon y Cajal de Madrid, Madrid, Spain

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Introduction: Pulmonary arterial hypertension (PAH) is a rare disease that affects the small pulmonary arteries, producing gradual obliteration of arterial lumen leading to the progressive increase in pulmonary vascular resistance and, ultimately, right ventricular failure and death. Idiopathic PH refers to patients without an identifiable underlying cause, although some of them may have a genetic cause. Our aim was to define the baseline characteristics, initial therapy and risk factors for survival of patients with 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 2007. 47 patients with idiopathic PAH were analyzed.

Results: The mean age at diagnosis was 4.5 years, and there was a similar proportion of both sexes. 64% of patients were in functional class I-II. The mean pulmonary arterial pressure was 47,3±16,3 mmHg, with mean pulmonary vascular resistance index of 12±8,3 WU·m^2. Initial therapy was oral monotherapy in 67%, prostanoids monotherapy in 11% and combination therapy with prostanoids in 9%. At the end of follow up, 55% of patients had received prostanoids. The mean follow-up time was 8 years. The survival rates (free of death or transplantation) was 91% (95% confidence interval (CI): 79–97%), 82% (95% CI: 67–91%) and 70% (95% CI: 52–82%) at 1, 5 and 10 years. Lung transplantation was performed in 7 patients. The main factors influencing survival, after adjustment for pulmonary vascular resistance, were functional class III-IV (Hazard Ratio (HR) 4.6, p=0.022, 95% CI: 1.3–15.3) and diagnosis under the age of 2 (HR 4, p=0.031, 95% CI: 1.1–14.4).

Conclusions: Idiopathic Pulmonary hypertension in pediatric age affects both sexes equally and begins at a young age. Prostanoids were used in 55% of patients. The main factors influencing survival, after adjustment for pulmonary vascular resistance, were functional class III-IV and diagnosis under the age of 2.