CHRONIC THROMBOEMBOLIC PULMONARY HYPERTENSION: DIFFERENCES AND SIMILARITIES WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION.

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Introduction: A pulmonary hypertension (PH) registry (REHAP) was undertaken to study epidemiology, clinical characteristics and survival of pulmonary arterial hypertension (PAH) and chronic-thromboembolic PH (CTEPH) in Spain. Aim: To analyze epidemiology, clinical characteristics, medical treatment (MT) and survival in CTEPH patients compared with idiopathic PAH (IPAH) patients.

Methods: Voluntary reporting of incident CTEPH and IPAH cases from 2007 to 2013 was evaluated. 321 CTEPH patients and 287 IPAH patients were analyzed. Clinical parameters, biomarkers (pro-BNP), 6-minutes walking test (6MWT) and hemodynamic variables were compared at baseline. Survival was studied by Kaplan-Meier test.

Results: Table 1 shows the main variables at baseline in CTEPH patients compared with IPAH patients

	CTEPH (n=321)	IPAH (n=287)	p-value
Age, years ± SD	63 ± 16	55 ± 19	<0.001
Men, n (%)	140 (44)	90 (31)	0.002
FC I-II, n (%)	97 (30)	101 (35)	ns
FC III-IV, n (%)	224 (70)	186 (65)	ns
proBNP, pg/mL ± SD	1573 ± 1767	1740 ± 2287	ns
6MWT, m ± SD	358 ± 119	388 ± 124	0.008

PaO_2, mmHg ± SD	61 ± 11	69 ± 16	<0.001
mPAP, mmHg ± SD	46 ± 12	52 ± 15	<0.001
CI, $L \cdot min^{-1} \cdot m^2$ ± SD	2.3 ± 0.6	2.4 ± 0.8	ns
PVR, Wood ± SD	10 ± 5.8	11.6 ± 6.5	0.002

At 31/DEC/2013 the prevalence of CTEPH and IPAH was 8.9 and 8.7 cases/million adult inhabitants (MAI), respectively. In 2013 the incidence of CTEPH and IPAH was 1.29 and 0.72 cases/MAI, respectively. Pulmonary endarterectomy were performed in 87 patients with CTEPH (27%) and 234 (73%) patients were treated with targeted PH therapy. All patients with IPAH were treated with targeted PH therapy: 49 (17%) with calcium channel blockers due to positive vascular reactivity test, 87 (30%) with phosphodieterase-5 inhibitors, 61 (21%) with endothelin antagonist receptors, 13 (5%) with prostanoids, 26 (9%) with oral combined therapy and 15 (5%) with other combined therapies. Survival at 1, 3 and 5 years from diagnosis for CTEPH vs IPAH was 93.7% vs 91.5%, 84.4% vs 75% and 69.5% vs 64.8%, respectively (p = 0.08).

Conclusions: Compared with IPAH patients, CTEPH patients are older, higher proportions of men, less distance in the 6MWT and worse hypoxemia, but better hemodynamic parameters. No differences in survival and prevalence.