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Title: Pulmonary hypertension in Portugal: First data from a nationwide registry

Rui 244 Baptista ruibaptista@gmail.com MD [1], José 242 Meireles zemeireles@gmail.com MD [2], Ana 406 Agapito anafagapito@gmail.com MD [3], Graça 407 Castro castro2406@gmail.com MD [1], António 408 Marinho da Silva marinhosilva1@gmail.com MD [1], Teresa 414 Shiang teresashiang@gmail.com MD [4], Ana 409 Oliveira ana.oliveirinha@gmail.com MD [4], Daniela 410 Ferreira danisaferreira@gmail.com MD [4], Fabienne 411 Gonçalves fabiennegoncalves@gmail.com MD [2], Susana 412 Robalo-Martins la63@sapo.pt MD [5], António 413 Nunes Diogo antonionunesdiogo@gmail.com MD [5] and Abilio 243 Reis abilioreis@me.com MD [2]. [1] Cardiology, Hospitais da Universidade de Coimbra, Coimbra, Portugal ; [2] Internal Medicine, Hospital Santo António, Porto, Portugal ; [3] Cardiology, Hospital Santa Marta, Lisboa, Portugal ; [4] Pneumology, Hospital Santos Silva, Vila Nova de Gaia, Portugal and [5] Cardiology, Hospital de Santa Maria, Lisboa, Portugal .

Body: Pulmonary arterial hypertension (PAH) is a rare disease that must be managed in specialized centers integrated in a national network. Availability of epidemiological national data is critical for planning and regulation of healthcare in this field. We conducted a prospective, observational and multicenter registry in 5 portuguese centers. Adults with PAH and chronic thromboembolic PH (CTEPH) confirmed by right heart cath (RHC) were included. 79 patients were enrolled; 46 (58.2%) classified as PAH and 33 (41.8%) as CTEPH. PAH patients had a mean age of 43.4±16.4 years and the f/m ratio 1.9:1. Idiopathic PAH was present in 17 (37%) patients, followed by connective tissue disease (n=12, 26%), congenital heart disease (n=10, 22%), portopulmonary (n=5, 11%), heritable (n=1, 2%) and other etiologies (n=1, 2%). At baseline most patients were WHO class III or IV (71%). Baseline RHC: elevated RAP (7.7±5.9 mmHg), mPAP (50.6±17.9 mmHg) and mean PVR (11.4±6.5 Wood U), with a low CI (2.7±1.1 L.min-1.m-2). At baseline patients were medicated with conventional therapies; at follow-up, most were on single (50%), double (28%) or triple (9%) combination of specific therapy. 1-year survival was 93.5%. CTEPH patients were older (60.0±12.5 years) and had higher RAP (11.0±5.2 mmHg, p = 0.015), but 1-year survival (93.9%) was similar to PAH patients. Five CTEPH patients underwent pulmonary endarterectomy. We estimated an annual incidence of 1.5 and 1.1 per million in PAH and CTEPH, respectively. Our report describes nationwide data on the diagnosis, management and clinical course of groups 1 and 4 PH patients. Clinical presentation, hemodynamics and survival are comparable with those reported on other national registries.