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Title: Incident pulmonary arterial hypertension in a Brazilian reference center: Results of a four-year registry

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Body: Background: There is an increasing interest in the worldwide characteristics of pulmonary arterial hypertension (PAH) patients although data from the southern hemisphere are still scarce. Objective: To describe baseline characteristics of incident PAH cases from a large Brazilian reference center. Methods: All consecutive patients diagnosed as PAH between 2008 and 2012 were included in the study. Baseline clinical, functional and hemodynamic data were collected for all patients. Results: A total of 171 patients were enrolled in the study (mean age of 46.8 ± 14.6 yo, female/male ratio of 3.3:1 and 45.9% in functional class III or IV). Connective tissue disease (CTD), idiopathic (IPAH), schistosomiasis and porto-pulmonary hypertension (PoPAH) accounted for 29, 25, 17 and 10% of all cases, respectively. Mean pulmonary artery pressure and cardiac output were 50.8 ± 19.2 mmHg and 4.6 ± 1.7 l/min, respectively, with mean six-minute walk distance of 420 ± 111 m and mean BNP levels of 212 ± 300 pg/ml. Female predominance was more evident in CTD and higher cardiac output was found in PoPAH. Furthermore, IPAH patients were younger than the other groups (mean age 40.7 ± 15.5 yo). The higher prevalence of CTD, as compared to previous registries, might be explained by the existence of a large rheumatology reference center in our University. Conclusions: Our results highlight the importance of regional registries in rare diseases for reflecting the existence of specific clinical conditions and health policies, such as schistosomiasis and the interaction with the rheumatology department, in order to appropriately design regional strategies for management of PAH.