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Title: Sarcoidosis associated pulmonary hypertension (SAPH) in the Netherlands

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Body: Background: The development of pulmonary hypertension (PH) is an important risk factor for early mortality in sarcoidosis. The prevalence is of SAPH in Europe is largely unknown. Aim: To investigate the prevalence of and clinical parameters associated with SAPH in the Netherlands. Methods: We analyzed clinical data and transthoracic echocardiograms (TTEs), made routinely at our PH/Interstitial Lung Disease clinic, from consecutive patients. Sarcoidosis patients with reliable echocardiography, without left heart dysfunction, were included. An estimated systolic pulmonary artery pressure (sPAP) > 50 mmHg was considered PH. Possible PH was defined as sPAP 37-50 mmHg or sPAP ≤ 36 mmHg with signs of right ventricular dysfunction. Results: From 139 sarcoidosis patients, 130 were included with stage 0 (11), I (46), II (39), III (7), IV (16). 5 patients (3,8%) had PH and 6 (4,6%) possible PH. Patients with PH were all women, predominantly black (4 out of 5) with stage IV disease (4 out of 5). 4 out of 6 patients with possible PH (3 white, 3 Asian) had stage I disease. TLCO % pred was lower in PH compared to no PH (median: 46,0 (18-47) vs 78,50 (29,0- 119) p < 0,01). FEV1 % pred was decreased in PH compared to no PH (median: 48,0 (36,0- 96,0) vs 82,0 (30,0- 131,0) p< 0,05). TLC was decreased in PH (median: 61,0 (51,0- 91,0) vs 82,50 (46,0- 116)), but not significantly different. In possible PH pulmonary function showed no significant difference versus no PH. Conclusions: SAPH is uncommon in the Netherlands and mainly found in black women with stage IV disease and associated with decreased lung function. Unexpectedly, suspicion of PH was found in some patients with stage I disease, warranting further investigation.