Prevalence of sarcoidosis-associated pulmonary hypertension: cumulative analysis of two PULSAR studies

To the Editor:

We have read with great interest the study by HUITEMA *et al.* [1] showing a pulmonary hypertension (PH) prevalence of around 3% in a large cohort of predominantly Caucasian sarcoidosis patients in a Dutch tertiary centre. According to the authors, a European Caucasian cohort has never been studied before this PULmonary hypertension in pulmonary SARcoidosis (PULSAR) study. In this context, in 2013 we reported another PULSAR study (PULmonary hypertension in SARcoidosis) in an albeit smaller cohort from a German tertiary centre [2]. Of interest, we showed very similar results, with a prevalence of precapillary PH of 3.6% with a similar study design in Caucasian sarcoidosis patients. Taken together, we think that it is very remarkable that both PULSAR studies confirm consistent prevalences and a cumulative analysis of both PULSAR trials would demonstrate sarcoidosis-associated pulmonary hypertension (SAPH) proven by right heart catheter in 15/510 patients and precapillary PH in 13/510 patients (table 1). The strength of both PULSAR studies is the prospective design and the invasive measurement of haemodynamics in most patients with suspicion of PH in transthoracic echocardiography; therefore, we would propose to use these algorithms to evaluate PH in sarcoidosis patients with unexplained dyspnoea. Echocardiography can show different signs of PH, before the definitive diagnosis for PH should be made by right heart catheterisation. Regarding the advanced stages with fibrosis in a relevant part of SAPH patients in both studies, we think that in these Caucasian cohorts the obliteration of the vascular bed by parenchymal fibrosis is most often the cause; however, the role of different factors, including pulmonary vascular compression due to adenopathy, granulomatous arteritis, pulmonary veno-occlusive disease, portopulmonary hypertension and postcapillary PH due to cardiac sarcoidosis, remains open. As we know that PH is a complication of advanced pulmonary sarcoidosis and it is associated with increasing morbidity and mortality, it is important to increase the awareness of SAPH and also to identify these different phenotypes.

TABLE 1 Cumulative analysis of two PULSAR studies showing prevalence of sarcoidosis-associated PH among sarcoidosis patients

Prevalence	PULSAR 2013 [2]	PULSAR 2019 [1]	Cumulative values
PH suspected by echocardiography	23/111 (20.7%)	28/399 (7%)	51/510 (10%)
PH proven by right heart catheterisation	5/111 (4.5%)	10/399 (2.5%)	15/510 (2.9%)
Precapillary PH	4/111 (3.6%)	9/399 (2.3%)	13/510 (2.5%)

PULSAR: PULmonary hypertension in (pulmonary) SARcoidosis; PH: pulmonary hypertension.

Treatment regimens were not assessed in both studies. Several drugs have been used to treat SAPH and have been investigated in different registries [3–6], showing that pulmonary arterial hypertension-specific treatment may be helpful in selected patients; however, controlled trials are rare. Next steps should be detailed phenotyping of SAPH patients and further studies that are needed to evaluate different treatment options in these selected SAPH phenotypes.

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A cumulative analysis of both PULSAR trials would demonstrate sarcoidosis-associated pulmonary hypertension proven by right heart catheter in 15/510 patients and precapillary PH in 13/510 patients
<http://bit.ly/34VGWbv>

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References

- 1 Huitema MP, Bakker ALM, Mager JJ, *et al.* Prevalence of pulmonary hypertension in pulmonary sarcoidosis: the first large European prospective study. *Eur Respir J* 2019; 54: 1900897.
- 2 Pabst S, Hammerstingl C, Grau N, *et al.* Pulmonary arterial hypertension in patients with sarcoidosis: the Pulsar single center experience. *Adv Exp Med Biol* 2013; 755: 299–305.
- 3 Skowasch D, Huscher D, Pizarro C, *et al.* Sarcoidosis-associated pulmonary hypertension *versus* idiopathic pulmonary arterial hypertension: results from the COMPERA registry. *Clin Res Cardiol* 2016; 105: Suppl. 1, P1798.
- 4 Boucly A, Cottin V, Nunes H, *et al.* Management and long-term outcomes of sarcoidosis-associated pulmonary hypertension. *Eur Respir J* 2017; 50: 1700465.
- 5 Baughman RP, Shlobin OA, Wells AU, *et al.* Clinical features of sarcoidosis associated pulmonary hypertension: results of a multi-national registry. *Respir Med* 2018; 139: 72–78.
- 6 Parikh KS, Dahhan T, Nicholl L, *et al.* Clinical features and outcomes of patients with sarcoidosis associated pulmonary hypertension. *Sci Rep* 2019; 9: 4061.

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