Management and long-term outcomes of sarcoidosis-associated pulmonary hypertension

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Abstract

Studies reporting the effects of modern strategies with pulmonary arterial hypertension (PAH)-targeted therapies in sarcoidosis-associated pulmonary hypertension (S-APH) are limited.

Clinical and haemodynamic data from newly diagnosed patients with severe S-APH (mean pulmonary artery pressure (mPAP) >35 mmHg or mPAP 25–35 mmHg with cardiac index <2.5 L·min−1·m−2) were collected from the French Pulmonary Hypertension Registry between 2004 and 2015.

Data from 126 patients with severe S-APH were analysed (mean±sd age 57.5±11.6 years, 74% radiological stage IV). 97 patients (77%) received PAH-targeted therapy and immunosuppressive therapy was initiated or escalated in 33 patients at the time of pulmonary hypertension diagnosis. Four months after PAH-targeted therapy initiation, mean±sd pulmonary vascular resistance decreased from 9.7±4.4 to 6.9±3.0 Wood units ($p<0.001$), without significant improvement in exercise capacity. Among the 11 patients treated only with immunosuppressive therapy, a haemodynamic improvement was observed in four patients, including two with compressive lymph nodes. After a median follow-up of 28 months, 39 patients needed PAH-targeted therapy escalation, nine underwent lung transplantation and 42 had died. Survival at 1, 3 and 5 years was 93%, 74% and 55%, respectively.

PAH-targeted therapy improved short-term pulmonary haemodynamics in severe S-APH without change in exercise capacity. Immunosuppressive therapy improved haemodynamics in selected patients. Pulmonary hypertension in sarcoidosis remains associated with a poor prognosis.