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Pulmonary arterial hypertension with below threshold pulmonary vascular resistance

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Selected patients with precapillary PH and "borderline" PVR, who fail to meet the current threshold of 3 Wood units, have functional limitation and adverse outcomes, and may potentially benefit from PAH therapy https://bit.ly/2QYaQrC

Cite this article as: Ratwatte S, Anderson J, Strange G, *et al*. Pulmonary arterial hypertension with below threshold pulmonary vascular resistance. *Eur Respir J* 2020; 56: 1901654 [https://doi.org/10.1183/13993003.01654-2019].

ABSTRACT Pulmonary vascular resistance (PVR) >3 Wood units is a criterion of the haemodynamic definition of pulmonary arterial hypertension (PAH). However, this cut-off is conservative and arbitrarily defined. Data is lacking on the natural history, response to therapy and survival of patients diagnosed with precapillary pulmonary hypertension (PH) with mild or borderline elevation of PVR.

In Australia, PAH therapy could be prescribed solely on mean pulmonary arterial pressure (PAP) and pulmonary arterial wedge pressure (PAWP) criteria. Using the Australian and New Zealand Pulmonary Hypertension Registry, we aimed to study a population diagnosed with PAH between January 2004 and December 2017 with the pre-defined haemodynamic characteristics of mean PAP ⩾25 mmHg, PAWP ⩽15 mmHg and PVR <3 Wood units.

Eighty-two patients met the pre-defined haemodynamic inclusion criteria (mean age 63±11 years; 67 females). Underlying aetiologies included idiopathic disease (n=39), connective tissue disease (CTD; n=42) and HIV infection (n=1). At diagnosis, mean PAP was 27 mmHg (interquartile range (IQR) 25–30 mmHg), PAWP 13 mmHg (IQR 11–14 mmHg) and PVR 2.2 Wood units (IQR 1.9–2.7 Wood units). Baseline 6-min walk distance (6MWD) was 352 m (IQR 280–416 m) and 77% of subjects were in New York Heart Association (NYHA) functional class 3 or 4. All patients were commenced on initial monotherapy with an endothelin receptor antagonist (ERA; n=66) or phosphodiesterase type-5 inhibitor (PDE5i; n=16). At first re-evaluation, 6MWD increased by 46 m (IQR 7–96 m) and 35% of subjects demonstrated improvement in

NYHA functional class. After a median follow-up of 65 months (IQR 32–101 months), 18 out of 82 subjects (22.0%) had died, with estimated 1-year and 5-year survival rates of 98% and 84%, respectively. Death attributed to PAH occurred in six out of these 18 patients (33.3%, 7% of total cohort).

Patients with precapillary PH and "borderline" PVR falling outside the current definition have adverse outcomes. Such patients appear to respond to PAH therapy; however, this requires further study in randomised trials.