



SHAREABLE PDF

EmPHasis-10 health-related quality of life score predicts outcomes in patients with idiopathic and connective tissue disease-associated pulmonary arterial hypertension: results from a UK multicentre study

Robert A. Lewis^{1,2}, Iain Armstrong¹, Carmel Bergbaum³, Melanie J. Brewis⁴, John Cannon⁵, Athanasios Charalampopoulos¹, A. Colin Church⁴, J. Gerry Coghlan⁶, Rachel J. Davies⁷, Konstantinos Dimopoulos³, Charlie Elliot¹, J. Simon R. Gibbs⁷, Wendy Gin-Sing⁷, Gulam Haji⁷, Abdul G. Hameed^{1,2}, Luke S. Howard⁷, Martin K. Johnson⁴, Aleksander Kempny³, David G. Kiely^{1,2}, Francesco Lo Giudice⁷, Colm McCabe³, Andrew J. Peacock⁴, Oyinkansola Peleyeju⁶, Joanna Pepke-Zaba⁵, Gary Polwarth⁵, Laura Price³, Ian Sabroe^{1,2}, Benjamin E. Schreiber⁶, Karen Sheares⁵, Dolores Taboada⁵, A.A. Roger Thompson^{1,2}, Mark R. Toshner⁵, Ivy Wanjiku⁶, S. John Wort³, Janelle Yorke⁸ and Robin Condliffe¹

Affiliations: ¹Sheffield Pulmonary Vascular Disease Unit, Royal Hallamshire Hospital, Sheffield, UK. ²Dept of Infection, Immunity and Cardiovascular Disease, University of Sheffield, Sheffield, UK. ³National Pulmonary Hypertension Service, Royal Brompton Hospital and Imperial College, London, UK. ⁴Scottish Pulmonary Vascular Unit, Golden Jubilee National Hospital, Glasgow, UK. ⁵Pulmonary Vascular Disease Unit, Royal Papworth Hospital, Cambridge, UK. ⁶Pulmonary Hypertension Unit, Royal Free Hospital, London, UK. ⁷National Pulmonary Hypertension Service, Hammersmith Hospital, London, UK. ⁸School of Nursing, Midwifery and Social Work, University of Manchester, Manchester, UK.

Correspondence: Robin Condliffe, Sheffield Pulmonary Vascular Disease Unit, Royal Hallamshire Hospital, Sheffield S10 2JF, UK. E-mail: robin.condliffe@nhs.net

@ERSpublications

The emPHasis-10 health-related quality of life score is an independent predictor of mortality in idiopathic and connective tissue disease-related pulmonary arterial hypertension, and has utility in risk stratification. <https://bit.ly/2MrLFLn>

Cite this article as: Lewis RA, Armstrong I, Bergbaum C, *et al.* EmPHasis-10 health-related quality of life score predicts outcomes in patients with idiopathic and connective tissue disease-associated pulmonary arterial hypertension: results from a UK multicentre study. *Eur Respir J* 2021; 57: 2000124 [<https://doi.org/10.1183/13993003.00124-2020>].

This single-page version can be shared freely online.

ABSTRACT Health-related quality of life (HRQoL) scores assess symptom burden in pulmonary arterial hypertension (PAH) but data regarding their role in prognostication and risk stratification are limited. We assessed these relationships using the emPHasis-10 HRQoL measure.

1745 patients with idiopathic PAH (IPAH), drug-induced PAH (DPAH), heritable PAH (HPAH) (collectively “(I/D/H)PAH”), or connective tissue disease-associated PAH (CTD-PAH), who had completed emPHasis-10 questionnaires at one of six UK referral centres between 2014 and 2017, were identified. Correlations with exercise capacity and World Health Organization (WHO) functional class were assessed, and exploratory risk stratification thresholds were tested.

Moderate correlations were seen between emPHasis-10 scores and 6-min walk distance ($r=-0.546$), incremental shuttle walk distance ($r=-0.504$) and WHO functional class ($r=0.497$) (all $p<0.0001$). Distribution of emPHasis-10 score differed significantly between each WHO functional class (all $p<0.0001$).

On multivariate analysis, emPHasis-10 score, but not WHO functional class, was an independent predictor of mortality. In a risk stratification approach, scores of 0–16, 17–33 and 34–50 identified incident patients with 1-year mortality of 5%, 10% and 23%, respectively. Survival of patients in WHO functional class III could be further stratified using an emPHasis-10 score ≥ 34 ($p < 0.01$). At follow-up, patients with improved emPHasis-10 scores had improved exercise capacity ($p < 0.0001$) and patients who transitioned between risk groups demonstrated similar survival to patients originally in those risk groups.

The emPHasis-10 score is an independent prognostic marker in patients with (I/D/H)PAH or CTD-PAH. It has utility in risk stratification in addition to currently used parameters. Improvement in emPHasis-10 score is associated with improved exercise capacity.