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Title: Severe pulmonary hypertension in patients with emphysema but preserved FEV1: Prognosis and response to treatment

Dr. Melanie 4451 Brewis mbrewis@nhs.net [1], Dr. Alistair 4452 Church colinchurch@nhs.net [1], Prof. Andrew 4453 Peacock apeacock@udcf.gla.ac.uk [1] and Dr. Martin 9535 Johnson mjohnson4@nhs.net [1]. [1] Scottish Pulmonary Vascular Unit, Golden Jubilee National Hospital, Glasgow, United Kingdom .

Body: Mild to moderate pulmonary hypertension (PH) is a common complication of chronic obstructive pulmonary disease. A smaller proportion present with severe PH with mean pulmonary artery pressures (mPAP) of >35mmHg. The role of PH therapies in this group is unknown. Methods:34 patients with CT evidence of emphysema but preserved FEV1(standardised residuals >-1.645) and mPAP >35mmHg at right heart catheterisation were diagnosed between 2000-2012. 54 idiopathic PAH (IPAH) patients from the same period were used for comparison. Six minute walk distance (6MWD), NTproBNP and functional class (Fc) were assessed at baseline and following 3 months of PH therapy. Results:In comparison to IPAH patients, there was no significant improvement in outcomes. (table1.) 1 and 3 year survival were worse in the emphysema group (Fig1). The strongest predictor of survival in the emphysema group was right atrial pressure (P<0.001). Conclusion. Compared with IPAH patients, patients with emphysema, preserved FEV1 and severe PH have worse survival and appear to respond poorly to PH targeted therapy.