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Title: Distinct clinical and pathological phenotypes in IPF

Dr. Elisabetta 23520 Balestro elisabetta_balestro@hotmail.com MD [1], Prof. Dr Fiorella 23521 Calabrese fiorella.calabrese@unipd.it MD [1], Prof. Dr Federico 23522 Rea federico.rea@unipd.it MD [1], Dr. Emanuela 23523 Rossi rossi.emanuela81@gmail.com MD [1], Dr. Francesca 23524 Lunardi francesca.lunardi@unipd.it [1], Dr. Marco 23525 Schiavon marco.schiavon@unipd.it MD [1], Dr. Erica 23526 Bazzan ericabazzan@hotmail.com [1], Dr. Monica 23527 Loy monica.loy@sanita.padova.it MD [1], Dr. Giuseppe 23528 Marulli giuseppe.marulli@unipd.it MD [1], Dr. Nazarena 23529 Nannini nazarena.nannini@unipd.it MD [1], Dr. Graziella 23530 Turato graziella.turato@unipd.it [1], Dr. Simonetta 23531 Baraldo simonetta.baraldo@unipd.it [1], Prof. Dr Manuel 23533 Cosio manuel.cosio@mcgill.ca MD [1] and Prof. Dr Marina 23549 Saetta marina.saetta@unipd.it MD [1]. [1] Department of Cardiothoracic and Vascular Sciences, University of Padova, Padova, Italy, 35127 .

Body: The clinical and functional course of IPF might deteriorate rapidly, rapid decliners (R) or slowly, slow decliners (S). Acute exacerbations (AE) are a feature of IPF. Lung pathology is considered to be similar in R and S; diffuse alveolar damage (DAD) is considered a common finding in AE. The aim of the study was a) determine the frequency of S and R phenotypes and incidence of AE in a IPF population followed before transplant, and b) correlate the clinical course with the quantitative lung pathology in the explanted lungs. 59 IPF patients referred for lung transplant were followed for 37 (12-156) months. A 10% FVC fall/year cutoff was used to define the R (>10%) and S (<10%)phenotypes. Lung pathology was quantitated in sections from upper and lower lobes. CD45 immunostaining was used to count inflammatory cells. 63% of patients were S and 37% were R. 74% of R and 73% of S were smokers. During the follow-up 23% of patients developed AE. The extension of lung abnormalities differed in the 3 groups; S had a significantly higher proportion of normal lung than R and AE (19 vs 9 vs 5% $p<0.05$), while R and AE had more intermediate and more honeycomb areas, respectively. DAD was present in 100% of R, 82% of AE and 12% of S ($p<0.005$) and vasculitis in 64% of R and 62% of AE but not in S ($p<0.005$). Severe cellular inflammatory infiltrate was seen in R and AE but not in S ($p<0.001$), and it was similar in smokers and nonsmokers. R and S decliners are different not only clinically but also pathologically and fulfill the definition of phenotypes. The degree of inflammation seen in R is the key finding separating the phenotypes. Smoking does not singly predispose for the R phenotype neither to the type and degree of lung inflammation.