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Title: Fibulin-1 is a novel biomarker of disease severity in pulmonary fibrosis

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Body: Idiopathic pulmonary fibrosis (IPF) is a severe form of interstitial lung disease (ILD) with a median survival post-diagnosis of 2-5 years. It is vital to distinguish IPF from other primarily inflammatory ILDs as the therapeutic options are distinct. Levels of the extracellular matrix protein fibulin-1 were evaluated in the serum and lung tissue from patients with ILD and levels correlated with lung function. Serum and pulmonary function tests from patients with ILD were collected from 3 cohorts (Sydney, Australia, Modena, Italy and San Francisco, USA). Fibulin-1 levels in serum were analysed by western blot. Immunohistochemistry was performed on tissue sections from people with no lung disease or IPF from 3 cohorts (Sydney, Perth, Australia and Modena, Italy) and staining intensity analysed by ImageJ. In ILD patients, serum fibulin-1 levels were associated with disease severity (composite physiologic index (CPI),DLCO% predicted) and elevated fibulin-1 levels predicted disease progression. In IPF patients, fibulin-1 serum levels were increased in patients with IPF compared to non-diseased controls (N=75 & 17; P<0.0001), and compared to non-IPF ILD patients (N=58; P<0.05). Fibulin-1 levels were increased in lung parenchyma (N=24; P<0.0001) in IPF patients compared to non-diseased controls, and were associated with disease severity (N=11; FEV1%: R=-0.78; P<0.005; FVC%: R=-0.75; P<0.01; CPI: R=0.64; P<0.05). Serum fibulin-1 is increased in patients with IPF compared to other ILDs and a high serum fibulin-1 level is suggestive of poorer survival. Parenchymal fibulin-1 correlates strongly with poorer lung function. Fibulin-1 may be an important prognostic molecule in ILD, particularly in IPF.