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Title: The clinical relevance of autoimmunity in idiopathic pulmonary fibrosis

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Body: The aim of this study is to evaluate the clinical significance of autoimmunity in IPF. This is a retrospective controlled study comparing clinical characteristics and outcome of IPF patients with (n=73) and without (n=87) positive autoimmunity. Seventy-three cases with positive autoimmunity included: 46 (63%) ANA (anti nuclear antibody), 20 (27%) RF (rheumatoid factor), 2 (3%) anti CCP (anti cyclic-citrullinated), 3 (4%) ENA (antibodies to extractable nuclear antigens), 2 (3%) ANCA (anti-neutrophil cytoplasmic antibody). No differences were found between patients with positive and negative autoimmunity in terms of age, gender, smoking history, prevalence of familial IPF, high resolution computed tomography features.

Characteristics of IPF patients.

	IPF-positive autoimmunity	IPF-negative autoimmunity
Age, median (range)	60 (47-9)	64 (39-84)
Male gender, n (%)	48 (66)	66 (76)
Smoking, current and former smokers, n (%)	58 (79)	59 (68)
Familial – IPF, n (%)	16 (22)	16 (18)
HRCT features consistent with UIP, n (%)	26 (36)	32 (37)
Median follow up, months (range)	32.19 (0.68-106.9)	32.48 (0.42-114.3)
AZA, CSS, NAC treatment, n (%)	9 (12)	15 (17)

During follow up two patients developed an autoimmune disease (one rheumatoid arthritis and one scleroderma). Survival between IPF patients with and without positive autoimmunity did not differ 77.4 months (95% CI 56.1-97.8). CONCLUSION A small minority (2.7 %) of IPF patients with positive

autoimmunity test at diagnosis developed an autoimmune disease during follow-up. The presence of positive autoimmunity testing do not influence neither IPF clinical presentation nor survival.