European Respiratory Society
Annual Congress 2012

Abstract Number: 3003

Publication Number: 2824

Abstract Group: 1.5. Diffuse Parenchymal Lung Disease

Keyword 1: Idiopathic pulmonary fibrosis **Keyword 2:** Interstitial lung disease **Keyword 3:** Interstitial lung disease (connective tissue disease)

Title: Significance of abnormal autoantibodies in patients presenting with IPF

Ms. Sam 8124 Hayward samhayward89@hotmail.co.uk [1], Dr. David 8126 McAllister david.mcallister@ed.ac.uk MD [2], Ms. Pauline 8127 Macfarlane p.mcfarlane@ed.ac.uk [1], Ms. Phoebe 8132 Wright phoebe-wright@hotmail.co.uk [1], Dr. Gareth 8134 Stewart gstewar1@staffmail.ed.ac.uk MD [4], Dr. William 8123 Wallace William.Wallace@luht.scot.nhs.uk MD [1], Dr. John 8125 Murchison John.Murchison@luht.scot.nhs.uk MD [1], Prof. John 8129 Simpson j.simpson@newcastle.ac.uk MD [3] and Dr. Nikhil 8122 Hirani n.hirani@ed.ac.uk MD [4]. [1] Department of Respiratory Medicine, University of Edinburgh, United Kingdom, EH17 4SA ; [2] Centre for Population Health Sciences, University of Edinburgh, Newcastle, United Kingdom ; [3] Institute of Cellular Medicine, Newcastle University, Newcastle, United Kingdom and [4] Centre for Inflammation Research, University of Edinburgh, United Kingdom .

Body: Introduction A subset of patients with IPF present with abnormal auto-antibodies (AAs) without clinical features sufficient to diagnose connective tissue disease (CTD). CTD-associated ILD is generally associated with a better prognosis than IPF. Aims To study the prevalence and significance of abnormal AAs in IPF. Methods Consecutively presenting patients with suspected IPF between 1/1/02 and 31/12/10 were prospectively recruited to a database. All IPF diagnoses required exclusion of overt CTD and an HRCT appearance of UIP with ≥ 70% probability. Patients with HRCT scans with ≥ 95% probability of UIP, or a confirmatory surgical lung biopsy were defined as definite IPF. The remainder were defined as probable IPF. Abnormal AA profile was defined as the presence of at least one of: RhF ≥40, ANA ≥1/640 and/or positive specific ENA screen. Patients were followed-up until Dec 2011 (median [IQR] among survivors 45 [25-63] months). Of 233 patients recruited, 25 did not have AAs performed within 12 months of presentation and were excluded. Of the 208 patients reported, 95 had definite IPF. Results AAs were abnormal in 18% of patients. Definite v probable IPF, gender, age, smoking and baseline lung function were similar for normal and abnormal AA groups. Only 3 patients developed overt CTD and all had abnormal AAs. Median survival was lower in those with abnormal AAs (39 v 69 months; unadjusted HR 1.57 [0.97 to 2.53] p=0.07; adjusted for age, sex, baseline VC, smoking and definite/probable IPF, HR 1.69 [1.03 to 2.78] p=0.04). Conclusions Only 1% of all IPF patients developed overt CTD. Abnormal AA serology was associated with a poorer survival.