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Title: Disease progression according to IPF phenotype

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Body: Introduction Our understanding of disease progression in IPF is based on cohorts of patients with 'definite' IPF. Such studies have reported a prognostic index for IPF (Du Bois 2011 AJRCCM 184; 459). In practice a diagnosis of definite IPF is not always attained; in some cases where the HRCT pattern is not incontrovertibly UIP, corroboratory invasive investigations are not performed. We define this condition 'probable IPF'. Aims To determine 1) disease progression and 2) the prognostic value of an IPF index in definite or probable IPF. Methods Consecutively presenting patients with IPF were prospectively recruited to a database. All IPF diagnoses required an HRCT appearance of UIP with ≥ 70% probability. Patients with HRCT scans with ≥ 95% probability of UIP, or a UIP biopsy were defined as definite IPF. BAL was not included in the diagnostic criteria. Results Of 193 patients, 89 had definite IPF. Median survival for definite v probable IPF was 3.2 v 6.0 years respectively (HR 1.45; 95%CI 0.94 to 2.25, p=0.09 adjusting for age, sex, height, VC and smoking). A relative decline in VC of 5-10% in the first 6 months was associated with increased risk of death (HR 3.12; 95% CI 1.55 to 6.30). Similar results were obtained in patients with definite and probable IPF. An IPF index was calculated in our cohort. Scores of 0-8 (n=97), 10-29 (n=76) and 30-61 (n=20) predicted median survival of 74, 48 (HR 2.02; 95%CI 1.26 to 3.23) and 12 months (HR 15.3; 95%CI 8.22 to 28.4) respectively ($p < 0.001$). The index had similar predictive value in definite and probable IPF. Conclusions Patients with both definite and probable IPF have poor prognoses but survival was worse in definite IPF. The IPF index may be of clinical value in definite and probable IPF.