TCO 62% (19). The median survival was 11.6 years, and all deaths (n = 12) were attributable to respiratory disease.

Summary Defining 'minimal' ILD is feasible and there was moderate radiological agreement. Minimal ILD is relatively benign, but the associated mortality was of respiratory cause.

S19 THE IMPACT OF CLOTTING ABNORMALITIES ON THE NATURAL HISTORY OF IDIOPATHIC PULMONARY FIBROSIS: AN EXTENDED FOLLOW UP OF A POPULATION BASED COHORT

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Background We have previously demonstrated that people with idiopathic pulmonary fibrosis (IPF) are more likely to have a prothrombotic state and that people with IPF and a prothrombotic state have a higher risk of death at a year's follow up. The aim of this study was to establish the impact of clotting abnormalities on the natural history of IPF with respect to median survival and lung function (forced vital capacity (FVC)).

Methods We recruited 211 incident cases of radiologically diagnosed definite or probable IPF and collected longitudinal information on pulmonary function tests done as part of routine care. All participants were tagged with the NHS Information Centre to enable us to collect data on mortality. Blood samples were tested for a prothrombotic state defined as at least one inherited or acquired clotting defect or marker of fibrinolytic dysfunction. Kaplan-Meier methods were used to calculate median survival. Random effects linear regression modelling was used to estimate decline in FVC.

Results Median follow-up was four years, during which 148 (70.1%) people died. Median survival in those with and without prothrombotic state was 2.7 and 3.7 years respectively (see Figure 1). We found evidence of effect modification between risk of death and follow-up time (p = 0.031). There was more than a three-fold increase risk of death in individuals with IPF and a prothrombotic state in the first half of follow-up (HR 3.36, 95% CI: 1.35 to 8.36), but this was reduced (HR 1.79, 95% CI: 1.08 to 2.94) in the second half. The estimated decline in FVC was 288mls (95%

Abstract S19 Figure 1 Kaplan-Meier survival estimates

CI: 184 to 392mls) in those with normal clotting and 328mls (95% CI: 269 to 387mls) in those with one or more clotting defects.

Conclusions Coagulation dysfunction has an adverse impact on the natural history of IPF, both in terms of median survival and lung function decline. Our findings suggest that a prothrombotic state may be a useful biomarker to predict prognosis as part of routine care.

S20 KBILD SCORES HAVE SIMILAR POWER TO PREDICT SURVIVAL AS PULMONARY PHYSIOLOGY IN INTERSTITIAL LUNG DISEASE

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Background The KBILD questionnaire is an ILD health related quality of life (HRQL) tool. Its relationship with survival has not been assessed.

Aims Assess impact of KBILD scores on survival in a heterogeneous population with interstitial lung disease (ILD).

Methods Patients attending the Bristol ILD service with fibrotic ILDs completed KBILD questionnaires, full lung function and exercise testing. Survival analysis using univariable and multivariable Weibull regression with an accelerated time-failure form was used to assess the significance of KBILD scores to predict all cause mortality. Comparison was made with lung function from the same clinic visit. Results are reported as hazard ratio and time ratio.

Area under receiver operator characteristics (AUROC) curve analysis was used to assess sensitivity of KBILD for predicting 12-month mortality.

Results 175 patients, 58% IPF, 67.4% male, completed a KBILD questionnaire. Mean values were; age 71yrs, KBILD 61, FVC

Abstract S20 Table 1 Weibull regression results and c-statistic for 12-month mortality for variables

	Weibull univariable regression		Weibull multivariable regression			AUROC	
	Hazard Ratio	Significance	Hazard Ratio	Time Ratio*	Significance	c-statistic	95% CI
Age (yrs)	1.05	0.003	1.06	0.96	0.001	0.646	0.511, 0.781
KBILD	0.98	0.005	0.98	1.01	0.022	0.654	0.531, 0.777
FVC (%)	0.97	<0.001	0.97	1.02	0.004	0.674	0.560, 0.788
Desaturation	2.64	0.002	2.00	0.61	0.038		
DLCO (%)	0.96	<0.001				0.680	0.554, 0.807
6MWD (m)	0.99	0.035				0.553	0.416, 0.690

TR – time ratio, AUROC – area under receiver operator characteristics curve, CI – confidence interval

Time ratio – The factor by which survival time changes for each 1 point change in a variable when all other variables are constant; eg. For each additional year of age, survival changes by a factor of 0.95