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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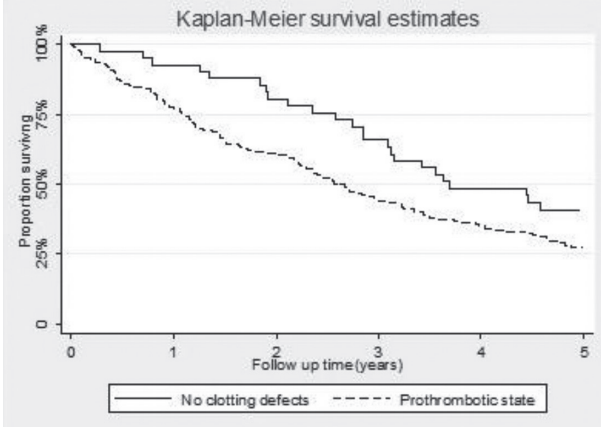
Background We have previously demonstrated that people with idiopathic pulmonary fibrosis (IPF) are more likely to have a prothrombotic state and that people with IPF and a prothrombotic state have a higher risk of death at a year's follow up. The aim of this study was to establish the impact of clotting 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-Meir 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% 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 prothombotic state may be a useful biomarker to predict prognosis as part of routine care.



Abstract S19 Figure 1 Kaplan-Meier survival estimates

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-statisitic	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

84%, DLCO 50%, walk distance 292m. 48% had oxygen desaturation on 6-minute walk. 47 patients (26.9%) died. Mean follow-up was 19.8 months, median 14.4 months. 156 patients had >12 months follow-up and these were included in the prognostic evaluation.

Univariable survival analysis showed age, KBILD, FVC, DLCO, walk distance and exertional desaturation to have prognostic significance for all cause mortality. Univariable analysis of the sub-categories of the KBILD score showed the psychological ($p = 0.003$) and breathlessness ($p = 0.002$) domains to be significant, while the chest symptoms domain was not ($p = 0.269$).

After backwards stepwise selection the multivariable model contained age, KBILD, FVC and desaturation (Table 1). All included variables had prognostic significance.

AUROC analysis showed KBILD had equivalent sensitivity for 12-month mortality to FVC, DLCO and better sensitivity than walk distance (c-statistic in Table 1). A KBILD score of 34 had 75% sensitivity for 12-month mortality, but only 10.5% specificity. Estimated median survival with KBILD of <34 was 9.7 months, compared to 36.4 months for KBILD > 34 ($p = 0.02$).

Conclusions In this cohort, the KBILD has equivalent prognostic power in ILD to pulmonary physiology and exercise testing at a single point in time. It is important to assess HRQL to give ILD patients optimal prognostic information.

S21 IDENTIFICATION OF CLINICAL PROGNOSTIC PARAMETERS IN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS

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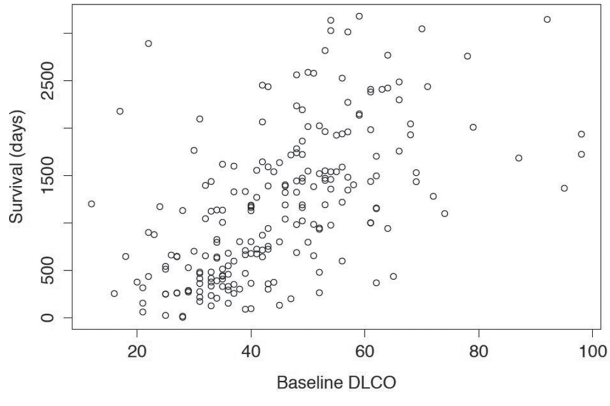
Idiopathic Pulmonary Fibrosis (IPF) is a progressive, scarring lung disease with a poor prognosis and median survival of 3 years. It is a heterogeneous disorder with varying rates of progression which presents a challenge for accurate prognostic prediction. The composite physiologic index (CPI) and the Gender, Age and Physiology (GAP) score are validated scoring systems for prognostic determination in IPF. Our data suggest these scoring systems have limited usefulness and we have undertaken a modelling approach to evaluate clinical prognostic parameters.

Methods Gender, age, smoking history, presence of emphysema on HRCT thorax and echocardiogram confirmed pulmonary hypertension were collected retrospectively from 253 IPF patients (in accordance with ATS/ERS criteria and MDT consensus) from a single centre in the UK between 19th April 2007 and 13th November 2014. Lung function including FEV_1, FVC, DLco and 6 minute walk test (distance, resting and minimum oxygen saturation and maximum heart rate) were collected at baseline, 6 and 12 months of follow up. Survival data were censored at 1st January 2016. The relationship between GAP or CPI and survival was analysed by Spearman's correlation, ROC area under the curve and Chi^2 analysis. Multivariate analysis and linear regression were used for the modelling.

Results Of the 253 patients included 188 were male (74%) with age 71.4 ± 8.3 years (mean ± SD). There were 164 (64.8%) ex-smokers and 12 (4.7%) current smokers. At presentation 19 patients had pulmonary hypertension and 35 had evidence of emphysema on HRCT thorax. Baseline lung function FEV_1 79 ± 22% predicted, FVC 82 ± 19% predicted, DLco 45 ± 15% predicted (mean ± SD). Median survival was 1169 days (3.2 years). The association between survival and CPI (r^2 0.59, $p < 0.01$) or GAP (r^2 0.45, $p < 0.01$) was modest. However ROC curve analysis demonstrated that GAP and CPI were poor predictors of survival. Chi^2 analysis shows there is no significant difference between these scoring systems. Multivariate analysis demonstrated that baseline% predicted DLco ($r^2 = 0.32$, $p = 6\times10^{-20}$) (Figure 1) had the strongest association with survival.

Conclusion Our data suggest that baseline percent predicted DLco may be a better predictor of outcome in patients with IPF. These results required validation by an independent cohort.



Abstract S21 Figure 1

Sleep Apnoea: The Big Sleep

S22 SEVERITY OF SLEEP DISORDERED BREATHING INDEPENDENTLY PREDICTS METABOLIC DYSFUNCTION IN A LARGE POPULATION OF SEVERELY OBESE SUBJECTS: THE ESADA STUDY

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Introduction Obstructive sleep apnoea (OSA) has an established independent association with insulin resistance and type 2 diabetes mellitus (T2DM). However, there are few data examining this relationship in severely obese populations, wherein any detrimental effect of OSA on metabolic health may conceivably be drowned out by the impact of morbid obesity. We assessed the relationship of OSA severity and nocturnal hypoxaemia with metabolic health in a cohort of severely obese patients attending sleep units across Europe.

Methods We performed a cross-sectional analysis of 1,434 participants in the European Sleep Apnea Cohort (ESADA) study with a body mass index (BMI) $\geq$35 kg/m^2, using multivariate regression analysis to assess T2DM prevalence according to OSA severity indices. Patients with diabetes were identified by history and medication prescription, and by screening for undiagnosed diabetes with glycosylated haemoglobin (HbA1c) measurement. The relationship of OSA severity with glycaemic control was assessed in diabetic subjects. Multivariate linear regression and multivariate analysis of co-variance were used to examine the