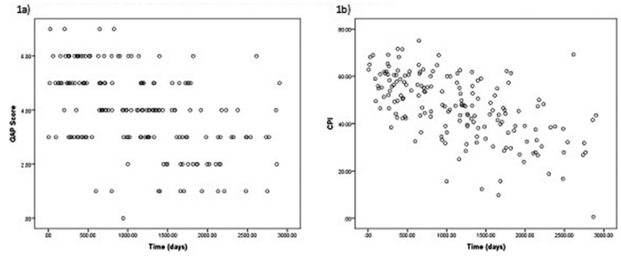
**Results** Of the 175 patients, 131 (75%) were male with a mean age of 71 ± 8 years (mean ± SD) at presentation. Overall 3-year mortality was 50%. The CPI demonstrated a better correlation with survival ($r^2$ = 0.37, p < 0.01) compared to the GAP score ($r^2$ = 0.24, p < 0.01). ROC curve analysis for 1-year mortality found that area under curve (AUC) was 0.726 for the GAP score and 0.783 for the CPI. For 3-year mortality AUC was 0.749 for the GAP score and 0.805 for the CPI.

**Conclusion** These data show that the CPI more accurately predicts survival at presentation and 3-years than the GAP score in a UK IPF cohort.




**Abstract S44 Figure 1** Correlation of GAP score (1a) and CPI (1b) and survival in IPF


**REFERENCES**

1 Wells AU, *et al*. *Am J Resp Crit Care Med*. 2003;**167**:962
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## S45 MUC5B GENOTYPE DOES NOT INFLUENCE COUGH SEVERITY IN IPF

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**Introduction** Cough is a disabling symptom in IPF which causes significant reduction in quality of life. The mechanisms underlying cough in fibrotic lung disease are not well understood. Polymorphisms in the rs35105950 promoter region of the *MUC5B* gene have been shown to relate to risk of developing IPF and, in those with disease, have been shown to increase mucin expression in the small airways. A previous small study linked carriage of the minor T allele at rs35105950 with heightened cough in IPF. We sought to test this observation in a larger cohort of prospectively recruited individuals with IPF.

**Method** *MUC5B* genotyping was performed on 117 prospectively recruited individuals with IPF. All subjects completed the Leicester Cough Questionnaire and full lung function at the time of their blood draw. Survival time from enrolment was censored at 1st May 2015. Statistical analyses were performed using SAS software.

**Results** The mean age of subjects at enrolment was 68 ± 8.4 with 79% being male. Subjects had, on average, moderately severe disease with mean FVC 73.9 ± 19.7% predicted and DLco 41.7 ± 18.1% predicted. At baseline, the median (IQR) LCQ score was 16.0 (11.6–18.6) points. 43 subjects (36.8%) were homozygous at rs35105950 for the G allele, 62 (53.0%) were heterozygous and 12 (11.1%) were homozygous for the T allele. Despite a trend towards worse disease at baseline, homozygosity of the T allele was correlated with a significant survival advantage (422 days compared to GG, p = 0.0063). There was, however, no difference on univariate analysis in LCQ cough severity between groups based on *MUC5B* genotype (GG 14.6 ± 4.5, GT 15.3 ± 3.9 and TT 15.1 ± 3.5, p = 0.70). This finding did not change when correction was applied for baseline disease severity.

**Conclusions** The severity of self-reported cough is worse in IPF than in other respiratory diseases. In contrast to one previous small study of 68 subjects, there is no relationship between patient reported cough and *MUC5B* genotype in our cohort. It therefore seems unlikely that the mechanism behind cough in fibrosing lung disease is related to mucin production. Further studies are required to determine the mechanisms underlying the heightened cough response observed in IPF.

## S46 AN RCT OF 28 DAY TREATMENT WITH FOSTAIR® PMDI 200/12 BD ON PLATELET BIOMARKERS IN PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS

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**Introduction and objectives** We have recently studied platelet activation in idiopathic pulmonary fibrosis (IPF) and found that IPF patients exhibit a significant increase in platelet reactivity. This was demonstrated by an ADP (Adenosine diphosphate) concentration dependent increase in platelet-monocyte aggregates (PMA), platelet P-selectin expression, and platelet fibrinogen binding.[1] We have suggested this may have a potentially important role in the initiation and/or progression of tissue injury in IPF.

Systemic corticosteroid treatment may alter platelet adhesion, as seen with suppression of p-selectin expression in the spontaneously hypertensive rat.[2] We hypothesised that peripherally deposited inhaled corticosteroid may have similar activity. In this study we evaluate the effect of beclomethasone/formoterol pMDI (B/F Fostair) on clinical parameters and biomarkers of platelet activation in IPF.

**Methods** Twenty non-smoking patients with IPF and no evidence of COPD were randomly assigned to either Fostair 100/6, 2puffs BD for 28 days or matched placebo inhaler. There was 28 days washout between crossover. Biomarkers, PMA, p-selectin and fibrinogen were measured at baseline and post treatment periods. Clinical outcomes of six minute walk (6MWT), spirometry, average daily activity over 7 days (Sense Wear) and Quality of life (KBILD) were also measured.

**Results** 17 patients (11 males, mean age 71.2 yrs) completed the study. Table 1 shows the 95% CI on differences between the baseline and two treatments on biomarkers of platelet adhesion obtained from ANOVA and using the Tukey *post hoc* test for multiple comparisons.

Change from baseline spirometric measurements of FEV1(L), FEV/FVC,% pred FEF25–75 were significantly improved following 28 days B/F by (mean ±SD), 0.88 ± 0.16 L (p = 0.03), 0.03 ± 0.03 (p = 0.03), 12.4 ± 19.1% (p = 0.02) respectively when compared to placebo.

There was no change in quality of life or exercise measures.