Idiopathic pulmonary fibrosis boldly goes where no disease has gone before

P1 PRELIMINARY RESULTS FOR ASSOCIATION OF SURVIVAL TIME IN IDIOPATHIC PULMONARY FIBROSIS CASES WITH THE 11P15.5 REGION

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Introduction Idiopathic pulmonary fibrosis (IPF) is a restrictive lung disease of unknown cause. The median survival time after diagnosis is 3–5 years and there are limited treatments available. *MUC5AC*, *MUC5B* and *TOLLIP* in the 11p15.5 region have been shown to be associated with susceptibility to IPF. A variant in *TOLLIP* (rs5743890) has also been shown to be associated with both susceptibility and survival time however the effects were in opposite directions, i.e. the allele associated with increased susceptibility was also associated with increased survival time.

Methods We performed survival analysis on 612 European IPF cases passing QC using a Cox proportional hazards model adjusting for age, sex, first 10 principal components and study centre. 134 variants were genotyped in a 200 kb region on chromosome 11 covering *MUC5AC*, *MUC5B* and *TOLLIP*.

Results No SNPs in this region reached genome-wide significance ($p < 5 \times 10^{-8}$). The most significant variant was rs56367042 (Hazard Ratio 3.38, 95% CI [1.91, 5.96]; $p = 2.74 \times 10^{-5}$) which is located downstream of *MUC5B*. The next significant SNP was rs5743894 (Hazard Ratio 0.67, 95% CI [0.54, 0.83]; $p = 2.24 \times 10^{-4}$) located in *TOLLIP*. For this SNP the allele we found associated with increased survival time has previously been reported as associated with increased susceptibility (showing a similar pattern to that reported for rs5743890) however this SNP has also been previously reported as not being associated with survival time. We found a proxy of rs5743890 ($R^2 = 0.698$) not to be associated with survival time ($p = 0.881$).

Conclusions Our results did not replicate those previously reported; however they may support the hypothesis that variants in *TOLLIP* that increase susceptibility may also increase survival time. In future we will perform a GWAS for susceptibility and survival time and try and replicate the findings.

P2 PLATELET REACTIVITY AS A POTENTIAL BIOMARKER IN IDIOPATHIC PULMONARY FIBROSIS

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Introduction and objectives Heterogeneity of idiopathic pulmonary fibrosis (IPF) means it is difficult to identify those at highest risk of progression who are most likely to benefit from treatment. A biomarker that predicts disease activity, prognosis and treatment response would be beneficial. We previously reported increased platelet reactivity in IPF,[1] and here we explore whether platelet reactivity warrants further investigation as a biomarker.

Methods Data were obtained from two studies: Study 1. a study of platelet reactivity in IPF; and Study 2. a pilot randomised-controlled trial of an investigational IPF treatment. Standard protocols were used to measure platelet-monocyte aggregate (PMA) formation, P-selectin expression, and fibrinogen binding in blood samples. Platelet reactivity in IPF was compared with controls. Correlation between platelet reactivity and forced vital capacity (FVC) was assessed. Study 2 data were used to assess the change in platelet reactivity in response to the intervention and correlation between baseline platelet reactivity, symptoms (KBILD) and exercise capacity (6MWD).

Results Study 1 included 13 IPF patients (mean± SD, Age 70.3 ± 5.8 years, 69% male, FVC 91.9 ± 17.8% predicted) and 12 controls (Age 66.2 ± 10.2, 66.7% males). Study 2 included 19 IPF patients (Age 71.5 ± 8.7, 72% males, FVC 84.4 ± 16.8% predicted). IPF patients demonstrated significantly increased platelet reactivity compared to controls ($P < 0.01$). Platelet reactivity and FVC did not correlate. In study 2, stimulated platelets expressed significantly less P-selectin in response to the intervention ($P = 0.03$). Unstimulated PMA formation moderately correlated with KBILD ($r = 0.38$. $P = 0.01$), but other platelet markers showed no correlation with symptoms or exercise capacity.

Conclusion IPF patients exhibit increased platelet reactivity compared to controls. The reduction in platelet reactivity in response to an intervention may indicate responsiveness to treatment effect. Although there was no correlation between FVC and platelet activation, further investigation is warranted to assess associations between platelet reactivity and lung function decline and mortality.

REFERENCE

1 Crooks MG, Fahim A, Naseem KM, Morice AH, Hart SP. Increased platelet reactivity in idiopathic pulmonary fibrosis is medicated by a plasma factor. *PLoS One* 2014;**9**(10):e111347

P3 PILOT STUDY TO TEST THE FEASIBILITY OF A PSYCHOLOGICAL SUPPORT WORKSHOP FOR PATIENTS NEWLY DIAGNOSED WITH IDIOPATHIC PULMONARY FIBROSIS (IPF) AND THEIR FAMILIES

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Introduction IPF is associated with a poor prognosis, high symptom burden and limited treatment options. Psychological reactions are comparable to those seen in cancer patients, yet support services for IPF patients are less well developed. There is an urgent demand to develop Interstitial Lung Disease (ILD) services that better support the psychological needs of IPF populations. The aim of this pilot study is to characterise the psychological needs of both patients and carers and to test the feasibility of a workshop approach.

Methods Consultant ILD physicians identified patients recently diagnosed with IPF at our Unit, and referred them to the clinical nurse specialist. Patients were invited to attend a half day workshop with an accompanying 'guest'. The workshop, led by a clinical psychologist, included didactic teaching, interactive discussions and experiential learning. Topics covered coping with low mood and anxiety, symptom control and transitions towards the end of life. Four members of the clinical team were present