P136 FACTORS INFLUENCING MORTALITY IN IPF PATIENTS TREATED WITH PIRFENIDONE

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Introduction and Objectives Idiopathic pulmonary fibrosis (IPF) is associated with a reduced life expectancy of 2–3 years from diagnosis to death. In the CAPACITY[1] study patients treated with pirfenidone had a slower decline in FVC (evident from 6 months), and showed a trend towards improved mortality. We investigated causes of death and factors influencing mortality in patients who have received pirfenidone on a Named Patient Programme.

Method We retrospectively analysed the data from 25 patients with a minimum of 3 months follow-up who were treated with Pirfenidone between August 2011 and June 2012 in a tertiary ILD clinic.

Results Mean age was 67±8 years and 19 (76%) were male. 8 of the 25 patients treated died. 4 had discontinued pirfenidone prior to death. 4 died due to an IPF exacerbation, 3 due to progressive disease and 1 due to a combination of progressive disease and congestive cardiac failure. Survivors had a greater DLCO % predicted at baseline (49±16 v 30±8; p=0.001) and were heavier (82.8±14.6kg v 71.0±11.2kg; p=0.032). A greater proportion of subjects who died (75%) had severe IPF (DLCO<35% predicted or FVC<50% predicted) compared with those who survived (18%; p=0.014). For survivors compared with those who had died, there was a trend towards greater BMI (28.7±4.4 v 25.4±3.9; p=0.065) and younger age (65±8.2 v 70±4.5; p=0.053).

FVC % predicted, Chronic Respiratory Questionnaire Scores and the presence of significant co-morbidities at baseline did not predict survival.

Conclusion In IPF patients who had been treated with pirfenidone, all died of IPF related causes, and those who died weighed less, and had more severe disease with a lower baseline DLCO% predicted.

This may help to inform us about the most appropriate patients for whom pirfenidone treatment should be considered.

1. Noble P, Albera C, Bradford W et al, for the CAPACITY Study Group. Pirfenidone in patients with idiopathic pulmonary fibrosis (CAPACITY): two randomised trials. Lancet 2011; 377: 1760–69.

P137 PRACTISE AND EFFECTS OF AUTOIMMUNE SCREENING IN PULMONARY FIBROSIS

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Introduction and Objectives Patients with pulmonary fibrosis can have features of autoimmune disorders that do not meet criteria for connective tissue disease (CTD), sometimes referred to as autoimmune-featured interstitial lung disease (AIF-ILD). Incorrect diagnosis and labelling could have important implications for treatment and prognosis.

We analysed the prevalence and type of immune profile performed to investigate patients with pulmonary fibrosis to determine what effect it had on subsequent management, and to assess variations in clinical practise.

Methods Retrospective review of patients attending the ILD and general respiratory clinics at our centre (n=75) with a diagnosis of pulmonary fibrosis, not known to be due to a CTD, or to be caused by a drug reaction, hypersensitivity, or sarcoidosis.

Results Seventy-five cases were reviewed (76% male). Age range was 55–88 years (median 73 years). Eighty percent of patients (n=60) had immune tests performed, with 13% (n=10) having two tests, 5% (n=4) having three, and 61% (n=46) having four or more. Twenty-eight percent (n=21) had a positive immune test. A positive immune screen partly directed the introduction of anti-inflammatory therapy in 24% of patients (n=5). One of these patients had a lung biopsy. In 38% of patients with a positive immune screen (n=7) it was unclear if the result contributed to clinical decision making. Lung biopsies were performed in 19% of patients with a positive immune screen, whereas only 8% of the total had a lung biopsy.

Conclusions These results show there is considerable variability in immune screen testing: which tests are performed, how positive tests are interpreted and whether they affect management. Our data hint at the usefulness of immune profiling, particularly where a lung biopsy has not been undertaken, and suggest that an abnormal immune profile increases the rate of lung biopsy. We suggest that a multi-centre, prospective, longitudinal study is required to further define AIF-ILD in terms of incidence, natural history, response to therapy and outcome. Whether AIF-ILD and the other idiopathic interstitial pneumonias are truly distinct remains to be determined, but clearly there is a need for advanced definitions, diagnostic algorithms, and management strategies.

Studies in PAH and pulmonary thromboembolism

P138 BASELINE HYPOXEMIA PREDICTS SURVIVAL IN PATIENTS WITH IDIOPATHIC PULMONARY ARTERIAL HYPERTENSION WITHOUT PATENT FORAMEN OVALE

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Introduction and Objectives The relevance of hypoxemia in patients with Idiopathic pulmonary arterial hypertension (IPAH) without Patent Foramen Ovale (PFO) on haemodynamics and survival is not well described. The aim of this study is to ascertain the severity and prevalence of hypoxemia in patients with IPAH without PFO, and its impact on survival.

Methods This is a single centre registry evaluation from 2001–2010 of patients attending the National Pulmonary Vascular Treatment Centre for Northern England. Outcomes were prospectively recorded on database following comprehensive assessment and confirmation with right heart catheterisation. PFO was excluded using bubble contrast ECHO. Data on arterial blood gas (ABG), lung function and radiology were obtained from case notes/hospital reporting systems. Patients initiated on targeted therapy were followed up at 3–4 month intervals.

Results ABG data was available for a cohort of 44 IPAH patients. 28(58%) were women. Mean age (SD) was 55.6(16.6) years, mean (SE) for Mean PAP 47(1.7) mm Hg, CI 2.18(0.09) L/min/m^2, 6MWD 266(22) metres, PaO_2 9.04(0.25) kPa, SPO_2% 93(0.3), FEV1% 81.1(2.8), DLCO% 61.4(3.4). Mild Hypoxemia was seen in7(16%), Moderate Hypoxemia in 26(59%), Severe Hypoxemia in 11 (25%) patients respectively. There were significant differences in survival between groups (logrank P value 0.0012, Figure 1). Mean survival (95% CI) in mild, moderate and severe hypoxemia groups were 12.3 years (9.1 to 15.4), 6.84(4.8 to 8.8), and 2.8(1.6 to 3.9) years respectively. There were no correlations between PaO2 and haemodynamic measurements (RAP/CI/PVR)/FEV1/FC. A modest correlation r. 32 was observed between PaO2 and 6MWD, (P0.04).

Conclusion Baseline Hypoxemia is an independent predictor of survival whilst on targeted therapy and moderate hypoxemia is prevalent in the absence of PFO/lung disease in IPAH.