pulmonary physicians to better understand practise patterns and unmet needs in IPF.

Methods In December 2010 – January 2011, a structured, quantitative survey was conducted with lung specialists in IPF and non-IPF specialist centres principally in five EU countries (Germany, Italy, Spain, UK, France [5EU]). The survey was extended to a further five countries where fewer physicians were targeted. The survey covered clinical practise patterns over the preceding 12 months. Data from the UK are presented here.

Results The 5EU sample included 232 participating physicians. Of these, 26 were from the UK, with 15 from specialist IPF centres, and 11 from general lung clinics. Among the UK clinics, 46% of patients had stable IPF, versus 56% with progressive disease (51% versus 49% respectively for 5EU). Triple therapy (steroid + immunosuppressant + N-acetylcysteine [NAC]) was prescribed for 31% of UK patients versus 25% for 5EU. Nineteen percent of UK IPF patients went untreated, and a further 10% received palliative care. Steroid monotherapy was used in another 10% of UK patients, and 11% received steroid + immunosuppressant. NAC + steroids were used in only 5% of patients, and NAC alone in 6%.

Conclusion IPF is a serious condition, but no standard therapy exists, and many patients receive no treatment or palliative care only. Current therapies are ineffective and unproven. At the time of the survey triple therapy remained prevalent; however, concerns have now been raised regarding AEs and risks of death and hospitalisation. There is therefore an unmet need for effective and well-tolerated therapies to enable a greater range of patients to be treated with agents that have been proven to be safe and effective in clinical trials.

P129 PATIENT AND PHYSICIAN DEMOGRAPHICS IN IDIOPATHIC PULMONARY FIBROSIS: RESULTS OF A UK PHYSICIAN SURVEY

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Introduction and Objectives Idiopathic pulmonary fibrosis (IPF) is a rare, progressive, fibrotic lung disorder of unknown origin, which leads to difficulty in breathing, functional disability and death. Due to symptomatic similarities with other lung conditions, IPF is difficult to diagnose, and patients experience varying standards of care. We undertook a survey of European pulmonologists to gain additional understanding of the characteristics of patients with IPF and the demographics of the physicians treating them.

Methods In December 2010–January 2011, a quantitative survey was conducted with lung specialists in IPF and non-IPF specialist centres in five principal EU countries (Germany, Italy, Spain, UK, France [5EU]). The survey was extended to a further five countries where fewer physicians were targeted. UK data are reported here.

Results The total number of patients seen by all the centres in the survey in the preceding 12 months was 7,302, 149 of which were from the UK. Among the UK patients, 87% were aged ≤85 years, which is similar to the value for 5EU (85%). Sixty percent of newly diagnosed UK patients were aged 65–80 years (5EU=56%), in contrast to 50% of the whole UK sample (5EU=57%). In 5EU, 70% of IPF patients were managed by 35% of pulmonologists, in contrast to the UK where 80% of patients were managed by 55% of pulmonologists. In the UK, 70% of pulmonologists were seeing fewer than 110 IPF patients annually, whereas in the 5EU, 70% of pulmonologists were seeing <30 IPF patients annually. Regional differences in the UK largely reflect population distribution with 14% of Scottish pulmonologists seeing ≥30 IPF patients, versus 33% for the north of England, 24% for the Midlands and Wales, and 29% for the south of England.

Conclusion In the UK, patient demographics for IPF were similar to the rest of Europe, but UK patients were being diagnosed at a younger age than previously compared to the rest of the EU. Importantly, the UK had a much wider spread of IPF patients among treating pulmonologists than the rest of Europe, which indicates a greater distribution of expertise in treating the disease.

P130 LIVING WITH IDIOPATHIC PULMONARY FIBROSIS: RESULTS OF A EUROPEAN PATIENT SURVEY

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Introduction and Objectives Idiopathic pulmonary fibrosis (IPF) is a rare, fibrotic, progressive lung disease that results in shortness of breath and difficulty in breathing. IPF commonly presents with crackling sounds in the lungs discernable via stethoscopic auscultation. IPF is most prevalent in patients >50 years of age. Risk factors include smoking and environmental exposures (metal, wood dust and other airborne pollutants). Disease progression features clubbing of the fingers, increasing difficulty in breathing, requirement for supplemental oxygen and/or lung transplantation. IPF is ultimately fatal. We conducted a European survey of diagnosed IPF patients to identify unmet needs in the management of IPF and opportunities to improve care.

Methods Patients in Germany, France, Italy, Spain and the UK with a confirmed diagnosis of IPF underwent a structured interview at home. In some instances, involved carers were also interviewed. The interviews featured associative questions where images were selected to express feelings, and constructive questions where factual recall was tested. Patient emotions were mapped according to four principle stages of the patient journey: 1. Symptoms, 2. Initial diagnosis, 3. After diagnosis, and 4. Disease management.

Results Forty-five patients (median age 67 years) and 18 care givers were interviewed for the survey. In 58% of cases, diagnosis was protracted due to dismissal of symptoms and misdiagnosis. Patients could be categorised in one of four ways (combative, serene, dejected and stoic) based chiefly on their age and attitude to disease management. Patients expressed a range of emotions at the point of diagnosis, and diagnosis was recognised as a time at which the patient-doctor relationship could break down. Patients treated in specialist centres reported better satisfaction with care than those treated by generalists. The patients cited negative impacts of IPF on almost all aspects of their lives, which could lead to depression. Keeping fit was acknowledged as a means to control disease symptoms and maintain a positive outlook.

Conclusion IPF has an enormous impact on patient quality of life, and patients are affected differently by the disease and its diagnosis. Emotional support and participation in patient support groups should be considered a key component of IPF management.

P131 WHAT IS THE EVIDENCE FOR PHARMACOLOGICAL AND NON-PHARMACOLOGICAL INTERVENTIONS IN IMPROVING DYSPNOEA, OTHER SYMPTOMS AND QUALITY OF LIFE IN PROGRESSIVE IDIOPATHIC FIBROTIC INTERSTITIAL LUNG DISEASE?- A SYSTEMATIC REVIEW OF THE LITERATURE

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Background Patients with Progressive Idiopathic Fibrotic Interstitial Lung Diseases (PIF-ILD) such as idiopathic pulmonary fibrosis have a short disease trajectory and have a similar prognosis to lung cancer patients. They have clear symptom control and quality of life (Qol) needs. The objective of this review was to evaluate the evidence for the use of pharmacological and non-pharmacological methods in improving dyspnoea, other symptoms and Qol for patients with PIF-ILD. In addition we assessed the use of outcome scales and economic evaluation of interventions.
Methods Studies were identified by searching eleven databases, relevant websites and hand searching key journals. Relevant studies were selected, assessed and data extracted independently by two researchers using standardised proformas. Meta-analyses were performed where appropriate and results presented as pooled mean difference with 95%CI. Effect sizes were also calculated where possible. A descriptive summary of other studies has been given.
Results 35 papers with 18 interventions were included. Meta-analyses were only possible for 3 interventions. Meta-analysis showed no significant treatment effect of IFN gamma 1b or sildenafil on 6MWD or dyspnea. A positive treatment effect of pulmonary rehabilitation on 6MWD (effect size (95% CI) 27.4 (4.1, 50.7) p=0.02) was seen. Separate analysis showed a positive effect of pulmonary rehabilitation on dyspnea and a trend towards significant results for pulmonary rehabilitation and sildenafil in improving quality of life. There was weak evidence for the improvement of 6MWD using oxygen, dyspnea using prednisolone, diamorphine, D-pencillamine and colchicine, cough using interferon alpha and thalidomide, anxiety using diamorphine, fatigue using pulmonary rehabilitation and Qol using thalidomide and doxycycline which warrants further research. There were a wide range of outcome scales used and no studies with economic evaluation.
Conclusion There is strong evidence for the use of pulmonary rehabilitation to improve 6MWD and moderate evidence for its use in improving dyspnoea and Qol. In addition, there is moderate evidence for sildenafil in improving Qol. There is weak evidence for a number of other interventions. Further research using economic evaluation and uniform outcome measures is needed.

P132 DEFINING BENCHMARKS FOR CLINICAL OUTCOMES IN IDIOPATHIC PULMONARY FIBROSIS

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Introduction and Oobjectives Idiopathic pulmonary fibrosis (IPF) is a rare, fatal, progressive, fibrotic lung disorder that results in reduced lung capacity and has a considerable deleterious effect on patient function. To date, there has been no consensus on the magnitude of treatment effect that constitutes a clinically meaningful response to IPF therapy. Since IPF shares a range of biological and prognostic features with non-small cell lung cancer (NSCLC), we conducted a systematic review of clinical trials evaluating the efficacy of therapies for NSCLC to establish a benchmark for the treatment of IPF.
Methods A literature search was performed to identify all randomised clinical trials between 1994–2010 evaluating therapies for NSCLC where a statistically significant effect of treatment on progression-free survival (PFS) or objective response rate (OR) was observed. The magnitude of the treatment effect in the NSCLC trials was compared to similar endpoints in three phase III clinical trials of pirfenidone in patients with IPF. In the NCSLC trials, PFS and OR were defined by standard conventions. In the IPF trials, PFS was defined as time to death or predefined thresholds for decline in forced vital capacity (FVC) or carbon monoxide diffusing capability. In the present analysis, objective response was defined according to predefined thresholds for change in FVC and the 6-minute walk test. Data were analysed according to the Cox proportional hazards model.
Results Twelve NCSLC trials, including a total of 13,959 patients, were identified by the search and included in the analysis. Of these studies, nine (12,456 patients) reported a significant effect on PFS, and seven (4,258 patients) reported a significant effect on OR. In both cases, the analysis showed that the magnitude of the response to therapy in the NSCLC trials was consistent with the pre-specified pirfenidone efficacy thresholds in the IPF trials.
Conclusion The clinical outcome parameters in therapeutic trials in NSCLC can be used to define benchmarks for assessing effect sizes in studies conducted in patients with IPF.

P133 A SINGLE CENTRE RETROSPECTIVE SURVIVAL ANALYSIS OF PATIENTS WITH INTERSTITIAL LUNG DISEASE

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Introduction Idiopathic pulmonary fibrosis is a chronic, progressive condition. BTS guidelines indicate that the 'triple therapy' - Prednisolone, Azathioprine, N-acetylcysteine (NAC) – can be considered in the management of UIP (usual interstitial pneumonitis) pattern disease based on the outcome of the IFIGENIA study (Demedts NEJM 2005). Recent data from the PANTHER study (Raghu NEJM 2012) has led to reversal of this recommendation due to concerns of increased adverse events, deaths and hospitalisation in the triple therapy arm as compared with placebo controls and those receiving NAC alone. This retrospective review examines the survival outcome of patients with UIP and UIP/COPD in a single centre (Sheffield Teaching Hospitals NHS Foundation Trust) where triple therapy was used over a five year period.
Methodology All patients with a UIP or UIP/COPD diagnosis managed within the interstitial lung disease clinic were identified through review of clinical notes and correspondence. Therapy, survival and demographic details were collected. Cox regression analysis was conducted using covariates of age at diagnosis, gender, occupational risk factors, COPD comorbidity and life limiting comorbidities.
Results 73 patients with UIP alone were identified; 16 patients had a UIP/COPD diagnosis. 8 patients received prednisolone alone, 2 NAC alone, 7 Pred/NAC, 3 Pred/Aza, 14 triple therapy and 11 received other immunosuppressive regimens (due to Azathioprine intolerance). 44 patients received no immunosuppressive therapy. Median survival, as derived from Kaplan-Meier curves, for those on no active therapy was 632 days (IQ range 485), 555 (IQ range 723) days for those on Prednisolone alone, 873 (IQ range 577)days for those on Pred/Aza and 869 (IQ range 918) days for those on triple therapy. No increase in hospitalisation was noted in the triple therapy group as compared with 'untreated' patients.
Conclusions In a cohort of 89 patients treated over a 5 year period triple therapy has been associated with an improved survival as compared to any other treatment regimen. No increase in hospitalisation has been identified and serious untoward events including blood abnormalities have been rare and manageable. Further Cox analysis using the covariates discussed above allied with smoking history, FVC and TLCO at diagnosis and rate of decline in FVC and TLCO at 'decision to treat' as covariates is awaited. We hope this data may enlighten practitioners to the progression of UIP over time, the reasons adopted by clinicians in selection of various (often unproven) therapies and the safety and utility of these treatments in a real life population.