airflow limitation (PAL)). There is no agreed definition of PAL in children, but pragmatically, most would define it as post steroid, post-bronchodilator FEV_1 percent predicted <80% or Z score less than −1.96, with normative data from appropriate reference populations despite optimal therapy. However, there is no agreement on the dose, duration or route of administration of steroids to determine the optimal spirometry that a child can achieve.

Hypothesis A single intramuscular dose of triamcinolone and acute bronchodilator are insufficient to determine optimal lung function and reliably diagnose PAL. The aim of this study was to determine whether forced expired volume in 1 second (FEV_1) 2–4 weeks after a single dose of triamcinolone and acute bronchodilator administration reflect the best obtained in the following year in patients with severe, therapy resistant asthma.

Patients and Methods 39 children age 5–16 received triamcinolone; the FEV_1 was measured before the treatment, after triamcinolone and during the 12-month period. The highest follow-up FEV_1 was compared with post-steroid post-post-bronchodilator FEV_1.

Results In the year following the 1st dose of triamcinolone 25 (64%) of 39 patients exceeded their immediate post-steroid trial target lung function by >9% predicted. 13 out of 39 patients (33.3%) achieved FEV_1 of >80% predicted at the 1st follow-up. If the diagnosis of PAL had been made just on the steroid trial, 16 patients would have been wrongly given this label; only 10 children were ultimately diagnosed with PAL. 13 of 39 patients received multiple (2–4) doses in 4 weeks intervals and only in 9 of them only the data was analysable. In this small group, the median and interquartile range of FEV_1 were significantly higher (75 vs 68 and 38.75 vs 17) following the 3rd dose of triamcinolone than after the 1st.

Conclusion Reliance on a single dose of triamcinolone plus acute administration of ß-2 agonist will lead to an overdiagnosis of PAL in children with severe asthma.

ILD: from bench to bedside and back again

P84 BTS NATIONAL INTERSTITIAL LUNG DISEASES (ILD) SURVEY 2010–2011

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Aim The aim of this survey was to capture the current "state of play" with regard to ILD services, with an emphasis on idiopathic pulmonary fibrosis (IPF), in the UK.

Methods The BTS Specialist Advisory Group emailed a survey to 260 clinicians. Respondents were advised to collect data prospectively for 1 month (late 2010/early 2011) before completing it.

Results 120 responses were obtained, (England-96, Scotland-16, Wales-6, Northern Ireland-2), total catchment population 37.3 million, number of full-time consultants 3 (1–15). *50% of centres had a "lead" ILD consultant, 57% ran an ILD clinic and, of those centres currently without an ILD clinic, 70% anticipated setting one up in the next 5 years. For patients with all types of ILD the estimated number of new patients seen at clinic each month was 6 (1–75). *The percentage of new patients with IPF (options "<25%", "25–50%", "50–75%" or ">75%") was estimated at 18%, 36%, 27% and 18% respectively. For all ILD patients, the number of return patients seen at clinic in a month was 25 (1–300). *Only 5% of respondents were "very confident" about these figures (based on audit/registry) with 62% expressing "low confidence". HRCT reporting was performed by "Pulmonary radiologists"/"Pulmonary radiologists with a specific ILD interest" in 48% and 36% respectively. 47% of centres did not have an ILD-multidisciplinary meeting (MDM), and for those that did, 32% held them monthly. Access to an ILD Specialist Nurse, ambulatory oxygen, pulmonary rehabilitation, palliative care and smoking cessation were 26% 93%, 81%, 93% and 97% respectively. 64% had not recruited into IPF clinical trials and 68% had no registry/database[n1]. "Triple" therapy continued to be prescribed "frequently", prednisolone (55%), azathioprine (49%), N-acetylcysteine (45%). The reported preferred model of care for ILD patients was "local" (51%), "network" (45%) and "centre" (4%). *Median/range.

Conclusions There is a wide variation in current practice, with almost half of all respondents not holding a MDM, despite BTS/ERS/ATS guidelines. Our survey has implications for the delivery of ILD care in the UK and should support the BTS in developing Quality of Care standards.

P85 MORTALITY TRENDS IN ASBESTOSIS, EXTRINSIC ALLERGIC ALVEOLITIS AND SARCOIDOSIS IN ENGLAND AND WALES

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Background To ascertain the trends in mortality from Asbestosis, Extrinsic Allergic Alveolitis (EAA) and sarcoidosis in England and Wales, we analysed mortality data from the Office of National Statistics.

Methods We calculated age and stratum specific mortality rates between 1968 and 2008 and applied these to the 2008 population demographics to generate the standardised number of expected deaths per annum. Poisson regression was used to calculate annual mortality rate ratios.

Results From 1968 to 2008 there were 1958 registered deaths from Asbestosis, 878 deaths from EAA and 3544 deaths from sarcoidosis. The Asbestosis mortality rate increased from 0.04 (95% CI 0.03 to 0.05) in the 1968 to 1972 calendar period to 0.12 (95% CI 0.10 to 0.13) in the 2005 to 2008 period while the mortality from EAA increased marginally from 0.04 (95% CI 0.03 to 0.05) in the 1968 to 1972 calendar period to 0.08 (95% CI 0.07 to 0.09) in the 2005 to 2008 period. Mortality from sarcoidosis has increased by approximately 9% a year.

Discussion Our findings show that the mortality from Asbestosis continues to rise in the UK. Overall mortality rates from EAA have remained stable throughout the same period but they were higher in males and in older people. There was a slight increase in mortality from sarcoidosis over the study period which was greater in women.

P86 THE ASSESSMENT OF HEALTH RELATED QUALITY OF LIFE IN SARCOIDOSIS WITH THE KING'S SARCOIDOSIS QUESTIONNAIRE (KSQ)

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Introduction The King's Sarcoidosis Questionnaire (KSQ) is a recently developed and validated sarcoidosis specific health related quality of life (HRQOL) tool comprising of 5 modules: general HRQOL (10 items), lung (6 items), medication/side-effects (3 items), skin (4 items), and eye (7 items). We set out to evaluate HRQOL in a large group of patients with wide ranging sarcoidosis and determine the factors that influence it.

Methods 207 patients with sarcoidosis (89% lung, 26% skin, 22% eye, 29% other organ involvement) attending outpatient clinics at King's College and Royal Brompton Hospitals completed the KSQ. KSQ domain scores range from 0 to 100, a higher score representing a better HRQOL. Demographic data, immunosuppressant medication, organ involvement, lung function, Scadding CXR stage, physicians global assessment (PGA) of severity of skin disease and visual acuity (VA) were recorded.

Results Patients had a mean (SEM) age 48 (11) years, 54% were female and 30% were Afro-Caribbean. Patients had a mean (SEM) FEV_1 80 (23)% predicted, FVC 94 (19)% predicted and TLCO % predicted 66 (17). HRQOL was impaired in all domains, mean (SEM) scores: general HRQOL 51 (2), lung 61 (2), medication/side-effects 49 (3), skin 54 (4), and eye 50 (4). Patients with 2 or more organ involvement compared to single organ involvement had worse general HRQOL (44 (3) vs 58 (3); $p<0.01$) and worse medication/side-effects scores (44 (3) vs 58 (3); $p=0.04$). Female patients compared to males had worse general HRQOL (45 (3) vs 57 (3); $p<0.01$) and medication/side-effects scores (41 (3) vs 58 (4); $p<0.01$). There were no associations between HRQOL and age ($r=-0.02$ to 0.13) or ethnicity ($p=0.42$). There was a weak but significant relationship between lung HRQOL and FEV_1 ($r=0.38$, $p<0.01$), FVC ($r=0.38$, $p<0.01$) and TLCO % predicted ($r=0.22$, $p<0.01$). Patients with Scadding CXR stage 3–4 disease compared to stage 0–2 disease had significantly worse lung HRQOL (51 (4) vs 63 (3); $p=0.02$). Skin health was associated with physician's global assessment (PGA) of severity of skin disease ($r=0.51$, $p<0.01$). Eye health was associated with VA ($r=-0.56$, $p<0.01$). Patients taking immunosuppressant medication for sarcoidosis compared to those not taking immunosuppresants had significantly worse general HRQOL (45 (2) vs 66 (4); $p<0.01$) and lung HRQOL scores (58 (3) vs 70 (4); $p=0.01$).

Conclusions HRQOL is impaired in sarcoidosis. Gender, immunosuppressant medication, multi-system organ involvement and severity of lung function impact on HRQOL. This study provides further clinical validation of the KSQ.

P87 THE NEEDS AND EXPERIENCES OF PROGRESSIVE IDIOPATHIC FIBROTIC INTERSTITIAL LUNG DISEASE PATIENTS, INFORMAL CAREGIVERS AND HEALTH PROFESSIONALS: A QUALITATIVE STUDY

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While there have been some studies looking at the needs of patients with idiopathic pulmonary fibrosis, to date no qualitative research has been conducted in the UK. This novel study aimed to assess the needs and experiences of people living with end stage progressive idiopathic fibrotic interstitial lung disease (PIF-ILD) and their informal caregivers. We also interviewed health professionals to examine views of current services, communication between health professionals and end of life planning. 18 qualitative semi-structured in-depth interviews were conducted with patients, their informal caregivers, and health professionals across two specialists ILD centres in London and in the community. Many participants reported that their main symptoms were shortness of breath and cough which impacted on every part of both theirs and the informal caregivers' lives. Psychologically, patients were frustrated and angry at the way in which their illness severely limited their ability to engage in activities of daily living and compromised their independence. Further, both patients and informal caregivers also reported that the disease seriously affected family relationships especially spousal relationships where strain was pronounced. Patients and their informal caregivers reported a good understanding of the progressive nature of their illness but held unrealistic expectations about prognosis. Health professionals expressed that there was a poor understanding of the palliative care needs of these patients, reluctance to recognise the terminal phase and poor end of life planning. All participants expressed that communication was often poor between health professionals and there was a lack of clarity about where primary responsibility for the care of the patient lay. This Phase I study has provided valuable insight into the overwhelming experiences of patients with PIF-ILD and their informal caregivers. It is clear that the disease affects the patients physically and also impacts greatly psychosocially. The palliative care needs of these patients are not being met and better co-ordination of care with improved communication is needed.

P88 THALIDOMIDE AS TREATMENT FOR IPF ASSOCIATED COUGH

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Introduction and Objective Idiopathic pulmonary fibrosis (IPF) is a chronic, irreversible fibrotic disease with more than 5000 incident cases annually in UK. IPF related cough is present in 80% cases and is often refractory to current treatments. Cough has a significant impact on quality of life, causing sleep disturbance, difficulties at work and stress incontinence. A previously published prospective open label phase II trial of thalidomide for treatment of pulmonary fibrosis suggested that IPF associated cough responded well to thalidomide. We have been using thalidomideas an "off-license" indication for selected IPF patients. The objective of this study was to review our experience of using Thalidomide as treatment for cough in IPF.

Methods Nine patients were referred to Nottingham Academic Interstitial Lung Disease clinic between 2009 and 2011 for assessment of their cough. A modified version of Leicester Cough Questionnaire was used, in conjunction with subjective symptoms, to clinically assess their cough. A trial of PPI (omeprazole 40 mg) and Prednisolone (10 mg) for 6 weeks was given to all subjects. Two were excluded as did not have significant cough, one patient declined thalidomide after initial screening.

Results Six patients were treated with thalidomide. Four had IPF, one had Hypersensitivity Pneumonitis and one had fibrotic Cryptogenic Organising Pneumonia. 72% were males with mean age 69 years (range 51–88 years). The median pre-thalidomide cough score was 74.5 (IQR 13.25) and post treatment cough score was 51.5 (IQR 49.25). This was statistically significant ($p=0.046$). Three stopped thalidomide subsequent to rash. Two patients are currently stable with 50 mg once daily, and 1 with 50 mg alternate daily of thalidomide.

Conclusion Our observation of a carefully selected cohort of patients suggests that thalidomide has potential for treatment of IPF associated cough. It works quickly (within days) as reflected both subjectively and objectively via questionnaire, even when prednisolone has failed. However, it does have a significant side-effect profile. Identification of thalidomide's mechanism of action may aid the development of novel, effective anti-tussive therapy for this debilitating aspect of IPF. We will assess this in randomised open label trial comparing thalidomide vs prednisolone for treatment of IPF associated cough.