Extrapolation from a North American population (ACCESS) may not be valid. This retrospective pilot study aimed to evaluate the potential of a novel web-based registry, by recording demographic and clinical characteristics, follow-up and treatment.

Methods 199 potential patients were identified from specialist respiratory clinics at two hospitals. 135 case notes were reviewed. Data were extracted from first and follow-up visits with random data quality checks by at least one other author.

Results Of 135 patients, 55% were female and 47% Afro-Caribbean; mean age (years) was 43.7±12.3 and patients had on average three follow-up visits per year. Presenting symptoms included cough (95%), breathlessness (41%), skin rashes (22%), arthralgia (15%), fatigue (19%), eye symptoms (14%) and fever (11%). 14% had stage 0 chest radiographs, 46% stage I, 16% stage II, 15% stage III and 9% stage IV. 45% had raised serum angiotensin-converting enzyme at first visit. 80% had histological confirmation. Systemic corticosteroids were the commonest treatment (21.5% after first visit). Cutaneous involvement was highest in females (64%) and Afro-Caribbeans (48%). Nodular lesions were the most common; 77% had granulomas on skin biopsy. 94% of those with skin disease had other organ involvement and received on average three treatments (topical 77%, prednisolone 64%, and hydroxychloroquine 45%). During follow-up, 73% reported improvement. 20 patients had definite/probable neurosarcoid (Zajicek criteria). Symptoms included parasthesiae (45%), visual disturbance (30%), headache (30%), seizures (10%) and anosmia (5%). Four had a VIIth nerve palsy. Diagnosis was made on MRI in 95%; 20% had CNS biopsies. 80% received prednisolone and 25% intravenous methylprednisolone. 15% were discharged after remission. Of the rest, 40% deteriorated during follow-up, 40% stabilised and 20% improved. Raised serum ACE was common in those with cutaneous and neurological disease.

Conclusions A novel web-based registry is a valuable method of recording clinical data in sarcoidosis. Additional use by other centres and the prospective recording of details of new cases could provide useful epidemiological data for the UK and drive future hypothesis-based studies.

S55 PULMONARY FIBROSIS WITH AND WITHOUT CO-EXISTING CONNECTIVE TISSUE DISEASE: DOES IT AFFECT SURVIVAL?

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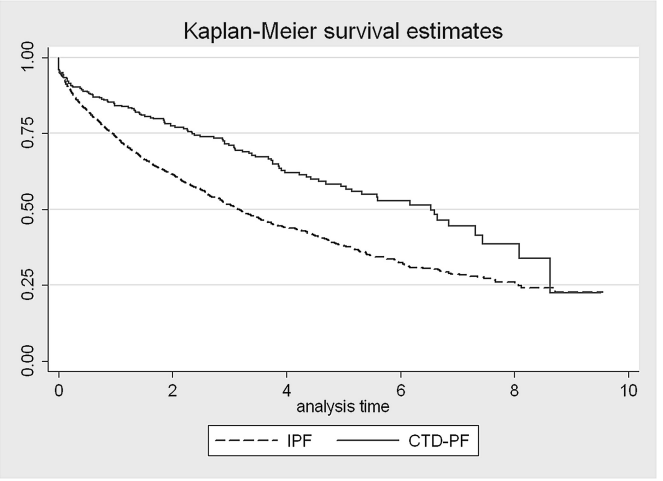
Introduction The impact of co-existing connective tissue disease on the survival of patients with pulmonary fibrosis remains unclear, since studies into the subject have so far yielded varying results. This study compares the survival of individuals with idiopathic pulmonary fibrosis (IPF) to those with connective tissue disease associated pulmonary fibrosis (CTD-PF) using data from The Health Improvement Network (THIN), a large general practice database in the UK.

Methods Incident cases of individuals with IPF and CTD-PF between the years 2000 and 2008 were identified. Survival analysis was performed on this dataset using Kaplan–Meier methods and Cox regression was used to compare mortality rates between patients with IPF and CTD-PF, adjusting for age and gender.

Results During the study period, there were 2208 incident cases of IPF, who were predominantly male (64%) and had a mean age at time of diagnosis of 73.9 years (95% CI 73.5 to 74.4 years). In comparison there were 318 incident cases of CTD-PF, of which the majority were female (56%) and the mean age at time of diagnosis was 67.8 years (95% CI 66.6 to 68.9). The commonest connective tissue disease associated with pulmonary fibrosis was rheumatoid arthritis (211 cases), followed by systemic sclerosis (39 cases). The mean follow-up period after diagnosis for our cohort was 2.4 person years and during this period 1117 (51%) patients with IPF and 116 (36%) patients with CTD-PF had died. The mortality rates for patients with IPF and CTD-PF were 221 (95% CI 208 to 234) and 121 (95% CI 101 to 146) deaths per 1000 person years respectively. The median survival for patients with CTD-PF was significantly higher at 6.5 years compared to 3.2 years in patients with IPF (p=0.0033) (See Abstract S55 Figure 1). After adjusting for age and gender, patients with IPF had a worse prognosis compared to those with CTD-PF (HR 1.32, 95% CI 1.08 to 1.60).



Abstract S55 Figure 1 Kaplan–Meier survival estimates.

Conclusion Individuals who have CTD-PF have a better prognosis after diagnosis when compared to those with IPF. This may relate to the differing natural histories of the diseases and needs further investigation.

S56 FLAT TRACHEA SYNDROME: AN UNDER-DIAGNOSED AND UNDER-TREATED CONDITION?

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Objectives Flat Trachea Syndrome (Tracheobronchomalacia) is a central airway disease characterised by weakness of the wall and dynamic decrease in the tracheal lumen and the large bronchi, particularly while exhaling. It causes chronic symptoms such as cough, dyspnoea, increase in recurrent infections, and poor secretion management, but it can also progress to chronic respiratory failure and death. It remains largely under-diagnosed unless clinicians are thoroughly acquainted with its peculiar symptomology being usually confused with other common diseases like chronic obstructive pulmonary disease (COPD) or asthma. Surgery with posterior tracheobronchial splinting (tracheobronchoplasty) using a polytetrafluoroethylene (PTFE) Teflon patch has been considered as a possible treatment option for this condition.

Methods A retrospective series of 28 patients that underwent tracheobronchoplasty using a PTFE Teflon patch between July 1998 and July 2010 were evaluated for symptoms, lung capacity and exercise capacity.

Results The age range of patients treated was from 24 to 82 (mean 66). 61% (17) of those treated were men, with all cases being diagnosed with obstructive airways disease on spirometry, with the FEV1 ranging from 34% to 65% pre-operatively. 25 patients (89%) presented with severe dyspnoea, 23 patients (82%) with uncontrollable cough, and 21 patients (75%) reported recurrent pulmonary infections. There was no in-hospital or 30-day mortality. After surgery symptomatic