

improvement in dyspnoea was reported in 24 of 28 patients ($p<0.001$), with the cough disappearing in 19 of 21 patients ($p<0.001$). At 3 months the FEV1 was $>80\%$ in all 28 patients. The mean exercise capacity was improved in 17 (61%) patients.

Conclusions The condition of tracheobronchomalacia is a misnomer and we propose the term “Flat Trachea syndrome” and/or “expiratory prolapse of the tracheobronchial posterior membrane”. It is a rare but severely debilitating condition which can be diagnosed easily by an awake flexible bronchoscopy and dynamic biphasic inspiratory/expiratory CT. Surgical airway splinting with a PTFE (Teflon) patch considerably improves respiratory symptoms, quality of life and functional status in highly selected patients with this under-diagnosed and under-treated condition.

S57 EVALUATION OF CIRCULATING BIOMARKERS IN LYMPHANGIOLEIOMYOMATOSIS

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Rationale Lymphangioleiomyomatosis (LAM) is a rare disease of the lungs and lymphatics occurring almost exclusively in women, usually presenting before menopause. It is characterised by progressive cystic destruction of the lung parenchyma, obstruction of lymphatics, airways, and often progressive respiratory failure. Recent diagnostic criteria require lung biopsy in addition to HRCT in the absence of other features such as tuberous sclerosis, renal angiomyolipomas or chylous effusions. The clinical course of LAM varies significantly, and there are no good predictors of clinical progression. Vascular endothelial growth factor D (VEGFD) has been found to be increased in the serum of LAM patients but its role as a biomarker has never been examined prospectively.

We aimed to see if:

1. VEGF-D reduces the need for lung biopsy for diagnosis using a proposed cut off with an estimated test sensitivity for LAM of 86%, specificity of 91%, and a positive likelihood ratio of 9.6.
2. VEGF-D is a useful predictor of severity when correlated with lung function data.

Results Serum samples were taken from 34 LAM patients and 12 healthy controls and a significant difference in VEGF-D levels was seen (Patient median=768 pg/ml, IQR=417.6–1509, control median=329.5, IQR=288.6–489.0, $p=0.0082$, Mann–Whitney U test). However, using the proposed cut off of 574 pg/ml by *Young et al* (2008), only 1 patient in our cohort with a diagnosis of “probable” LAM would have avoided the need for a lung biopsy to confirm diagnosis. When correlated with lung function, only TLCO demonstrated a statistically significant negative correlation with VEGFD levels ($r^2=0.2143$, $p=0.0131$).

Conclusions Though an interesting research tool, the value of VEGFD as a biomarker in LAM has not been clearly demonstrated and there is currently insufficient evidence to advocate its role either to aid in diagnosis or prediction of outcome in LAM.

S58 PULMONARY LANGERHANS' CELL HISTIOCYTOSIS (PLCH): A NEW NATIONAL REGISTER

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Introduction and Objectives PLCH is a rare interstitial lung disease, linked to cigarette smoking and, in some cases associated

with respiratory failure and death. Limited UK data have been published and hence little knowledge exists of the diagnostic and treatment practices employed by UK Respiratory physicians. Our study aims to characterise the epidemiological, clinical, histological, radiological and prognostic indicators in a nation wide cohort of patients with PLCH. This study aims to follow the patient journey from point of diagnosis, regardless of the mode of diagnosis, looking at patient demographics and clinical outcomes in the UK population.

Methods UK Respiratory Consultants were contacted to request details for 71 patients with known PLCH, who had previously been registered on the BTS Orphan Lung Disease (BOLD) database. Patients were sent a consent form and questionnaire. Once written consent was obtained, consultants were sent a questionnaire requesting medical information. The patients' GPs have provided current medication and past medical history information.

Results Details on 55 patients were received (including 10 deceased and, 4 lost to follow-up). 17 patients (8 males), have returned a completed consent form and questionnaire so far, and were included for analysis. Age at presentation 32.0 years (SD13.7). *Presenting symptoms:* Shortness of breath 70.5%, pain 47.1%, pneumothorax 11.8%, cough 29.4% and 5.9% asymptomatic (diagnosed incidentally). *Smoking status:* Ex 88.2%, current 6.0% mean (SD) 22 (27.2) pack years, and 6.0% never smokers. 23.5% admitted limited exposure to cannabis. *Diagnosis:* 82% patients had had an HRCT scan and 64.7% an open lung biopsy. *Diseasecourse:* Symptoms resolved 35.3%, same 41.2% and 23.5% had slowly progressed. 47.0% pneumothorax, 53.0% have received treatment either chemotherapy 29.4% (Chlorambucil (2), 2-Chlorodeoxyadenosine (2), Azathioprine (1)) or, an operation for recurrent pneumothorax (23.5%) 1 patient is on the waiting list for a lung transplant. *Patients' opinion of Doctors' knowledge of PLCH:* High 35.3%, medium 23.5%, Med/low or low 41.1%.

Conclusions Although small, this early dataset indicates a high prevalence of smoking in our UK cohort and, that despite advances in CT a high percentage of patients are still diagnosed with an open lung biopsy.

α 1-Antitrypsin: what it tells us about COPD

S59 α -VAL360: A PLASMA MARKER OF NEUTROPHIL ELASTASE ACTIVITY AND COPD DISEASE SEVERITY

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In both A1AT deficient and replete subjects, it is widely believed that protease-antiprotease imbalance is central to the pathophysiology of COPD,¹ although (in the absence of an exacerbation) it is difficult to detect free elastase activity.² This is explained by recent mathematical and in vitro modelling suggesting proteolysis and subsequent enzyme inhibition occur within the immediate micro-environment of the neutrophil.³ We have therefore validated a (pre-inhibition) elastase cleavage product of fibrinogen (α -Val³⁶⁰) produced in this microenvironment as a potential marker for COPD.

Methods *Pilot study:* Plasma α -Val³⁶⁰ was measured in 68 subjects with a wide range of A1AT levels, and in a further 64 PiZ A1AT deficient subjects, spirometry and plasma α -Val³⁶⁰ were measured in the stable state. *Subjects with COPD (Normal A1AT levels):* 81 subjects were recruited in a primary care setting following an exacerbation associated with sputum production. Trial participants were assessed on day 1 of the exacerbation and in the stable clinical state when full lung function tests and HRCT were also performed. HRCT scans were analysed both densitometrically (voxel index) and visually.