Non-IPF ILDs: diagnosis and management

P28 REAL WORLD MDT DIAGNOSIS OF IDIOPATHIC PULMONARY FIBROSIS

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Idiopathic pulmonary fibrosis (IPF) is the commonest idiopathic interstitial lung disease (ILD). The diagnosis is determined by a usual interstitial pneumonitis (UIP) pattern on high resolution computed tomography (HRCT). Current guidelines advocate lung biopsy in patients with diagnostic uncertainty though biopsy can afford significant morbidity and mortality. We aimed to evaluate our practice in diagnosing IPF in relation to these guidelines.

Methods We evaluated our experiences in a multidisciplinary team (MDT) setting in a UK tertiary referral centre of 104 patients referred with a presumed diagnosis of IPF between November 2012 and July 2014.

Results After MDT discussion, 48.5% patients had definite UIP and 51.4% had possible UIP or fibrotic non-specific interstitial pneumonitis (NSIP) based on ATS/ERS criteria. Of the fifty-three patients with possible UIP/NSIP, fifteen (28%) patients had a lung biopsy. Twelve out of fifteen patients had UIP on biopsy (80%). One patient died and one suffered with chronic pain post-biopsy (13%). In the remaining thirty-eight (72%) patients, biopsy was not possible due to comorbidities or patient choice.

Of the thirty-eight patients with radiological diagnosis of possible UIP/NSIP, thirteen (34%) patients were deemed to have a clinical diagnosis of probable IPF after MDT discussion based on disease progression and age. Two (5%) patients were subsequently diagnosed as having a connective tissue disease and twenty-three (60%) patients were clinically diagnosed as NSIP based on response to immunosuppression and stability of lung function.

Conclusions Surgical lung biopsy is considered the gold standard within the diagnostic work-up when there is diagnostic uncertainty. However in our clinical practice over two thirds of patients are not suitable for biopsy due to comorbidities and patient choice. Our clinical experience of a high yield of UIP after biopsy in patients with a radiological diagnosis of possible UIP/NSIP and concerns regarding morbidity and mortality has altered our practice. We increasingly utilise clinical data regarding progression and failed response to immunosuppressive therapies to aid in the MDT diagnosis of IPF when there is diagnostic uncertainty. In the subsequent year, 8.7% of patients underwent biopsy versus 14.6%, reflecting a change in practice.

P29 BRISTOL INTERSTITIAL LUNG DISEASE (BILD) SERVICE EXPERIENCE: BILDING ON THE MDT

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Introduction The weekly Bristol Interstitial Lung Disease (BILD) multidisciplinary team (MDT) meeting decides consensus diagnoses for patients from around the South West. A database records cases at the time of discussion. Referrals have increased since the advent of novel agents for Idiopathic Pulmonary Fibrosis, prompting this retrospective review of the MDT experience.

Aims Establish the range of cases referred, determining the proportion for whom MDT discussion leads to changes in diagnosis and which variables influence this. Examine IPF patients, identifying differences between those prescribed Pirfenidone or otherwise.

Methods For all cases recorded in the MDT database between 1/1/2013 and 1/1/2015, the pre-MDT differential diagnosis and consensus diagnosis, dates of referral/discussion, referral source, demographics, investigation results, the number of discussions and dispensing of Pirfenidone were reviewed. For patients with multiple entries, initial differential diagnoses were compared to final consensus. Outcome measures of interest were change in diagnosis and decision to use Pirfenidone in IPF patients.

Results 846 discussions occurred (651 individual patients) over this period. Pre/post MDT diagnoses are shown in the Table 1. 78% of cases were discussed within 2 weeks of referral. 25% were discussed more than once (range 1–5). 54.7% of IPF cases were external referrals vs 32.3% overall.

Diagnosis changed following discussion for 44.1% of patients. Pre-MDT diagnosis of IPF changed for 36.7%. Logistic regression suggests pre-MDT differential diagnosis and age at referral are main influences on change in diagnosis.

Overall mean age was 65.5 years (17–91), 58.1% male. For IPF cases, mean age was 74.4 years, 76.9% male. Pirfenidone was prescribed to 46.2% of IPF cases; median time to dispensing 61 days. 6MWD was greater where Pirfenidone was given (284 m vs 249 m, p = 0.03); however lung function and HRCT pattern did not differ. 12-month mortality was 6.7% in the Pirfenidone group, 27.1% where not given (p = 0.002).

Abstract P29 Table 1 Pre-MDT diagnoses and consensus diagnoses following MDT discussion

Diagnosis	Pre-MDT Diagnosis		Consensus diagnosis		1 year transplant free survival
	n	%	n	%	%
Asbestosis	10	1.5	11	1.7	81.8
CPFE	26	4.0	33	5.1	78.8
CT-ILD	79	12.1	68	10.4	98.5
Drug Related ILD	16	2.5	14	2.2	92.9
Hypersensitivity pneumonitis	63	9.7	71	10.9	85.9
IPF	150	23.0	130	20.0	81.5
No ILD	2	0.3	65	10.0	89.2
NSIP	47	7.2	73	11.2	84.9
NSIP/UIP Spectrum	12	1.8	6	.9	83.3
Organising pneumonia	11	1.7	11	1.7	90.9
Other (including vasculitis, DIP, LAM etc)	33	5.1	30	4.6	90
Pulmonary Langerhans Cell Histiocytosis	5	0.8	7	1.1	100
RB-ILD	10	1.5	13	2.0	100
Sarcoidosis	84	12.9	74	11.4	97.3
Unclassifiable ILD	103	15.8	45	6.9	84.3
Total	651	100.0	651	100.0	88.2

CPFE: Combined Pulmonary Fibrosis/Emphysema, CT-ILD: Connective Tissue disease-associated ILD, IPF: Idiopathic Pulmonary Fibrosis, NSIP: Non-specific Interstitial Pneumonia, UIP: Usual Interstitial Pneumonia, RB-ILD: Respiratory Bronchiolitis-ILD, DIP: Desquamative Interstitial Pneumonia, LAM: Lymphangioleiomyomatosis

Conclusion Specialist MDT discussion influenced changes in diagnosis in 44.2% of patients. The majority of IPF cases discussed are external referrals. This has implications for design and delivery of specialist ILD services.

Case selection for Pirfenidone does not appear based on lung function differences. This has implications for guidelines for its use.

P30 EFFICACY OF PULSED CYCLOPHOSPHAMIDE AND METHYL-PREDNISOLONE THERAPY IN PATIENTS WITH PROGRESSIVE INTERSTITIAL LUNG DISEASE

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Introduction Therapeutic options for progressive interstitial lung disease (ILD) are limited, as no pharmacological intervention has been shown to improve mortality significantly. Intravenous (IV) pulsed cyclophosphamide and methyl-prednisolone are administered in some centres for rapidly progressive ILD, based on small clinical improvements in patients with ILD secondary to systemic sclerosis. We studied the outcome of patients with ILD who received IV cyclophosphamide and methyl-prednisolone therapy in our centre.

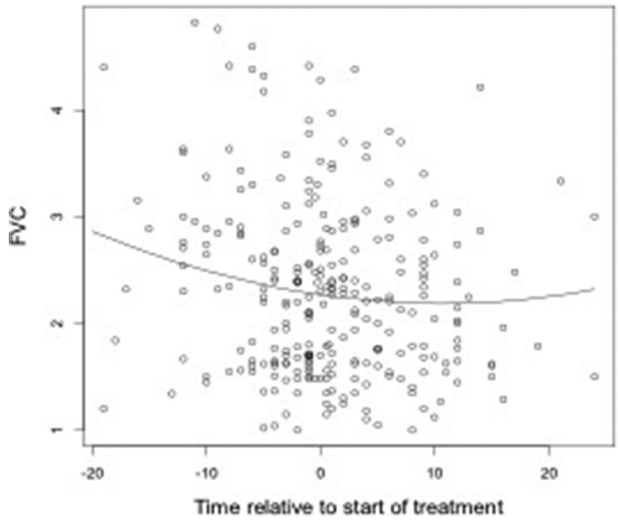
Methods Patients were identified from a database of those receiving cyclophosphamide. We reviewed the case notes of patients receiving IV cyclophosphamide between January 2010 to August 2014, comparing the rate of change in forced vital capacity (FVC) and transfer factor for carbon monoxide (TLco) before and after therapy. Adverse events were also recorded.

Results Records from 53 patients with a mean age of 60 years (range 39 – 81 years) were reviewed; 29 (55%) were male. Diagnosis included Connective Tissue Related-ILD (21), Idiopathic NSIP (12), Chronic Hypersensitivity Pneumonitis (8), IPF (6) and Undifferentiated-ILD (6). The median number of cyclophosphamide pulses received was 6 (range 1 to 23). After completion of cyclophosphamide, 41 (77%) patients received follow on immunosuppressive therapy in the form of mycophenolate mofetil (37 patients), azathioprine or rituximab.

The average rate of change of lung function was significantly less after cyclophosphamide therapy both for FVC (Figure 1) $p = 0.0004$ and TLco $p = 0.00015$.

Whilst on therapy 8 (15%) patients had bone marrow suppression (new onset neutrophil count $<2.0 \times 10^9$/l and/or platelet count $<140 \times 10^9$/l), 4 (7%) had elevated liver function tests, 3 (6%) had abnormal renal function, and one was investigated for microscopic haematuria. 32 episodes of infections were documented of which 21 were of respiratory origin. 25 (71%) out of 35 patients who suffered from an adverse event were able to complete therapy.

Conclusion In our single centre, retrospective study, pulsed intravenous cyclophosphamide and methyl-prednisolone was associated with stabilisation of lung function in a mixed cohort of patients with progressive ILD. Adverse events were common but transient and managed with dose reduction and/or delayed schedule.



Abstract P30 Figure 1 Showing data plots and average fitted quadratic curve for FVC of patients who received IV Cyclophosphamide therapy. FVC (litres); Time relative to treatment (months)

P31 A RETROSPECTIVE ANALYSIS OF INTERSTITIAL LUNG DISEASE SCREENING IN A REGIONAL CENTRE FOR PATIENTS WITH SCLERODERMA

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Background Interstitial Lung Disease (ILD) and Pulmonary Arterial Hypertension (PAH) are the major sources of morbidity and mortality amongst patients with Scleroderma. Specific autoantibodies, anti-Scl70 and anti-centromere (ACA), are associated with ILD and PAH respectively. Screening for ILD and PAH using annual pulmonary function testing (PFT), High Resolution Computed Tomography (HRCT) and Echocardiography respectively, is recommended by the BTS ILD Guidelines, 2008. However, the predictive value of autoantibodies and clinical screening for ILD and PAH, remains unclear in regional centres managing patients with Scleroderma. We hypothesised that an objective scoring system would elucidate lung phenotypes amongst the cohort and confirm original radiology reports for these patients, whilst patients autoantibody profiles would serve a clinical purpose in management. We retrospectively compared identification of ILD by a specialist ILD radiologist, against the use of predictive autoantibody profiling, for the detection of ILD.

Methods 99 patients with Scleroderma, managed in Nottingham, were identified from clinic lists (n = 68) or the pathology database, for positive anti-Scl70 and ACA results (n = 31). Autoantibody profiles (n = 77), including Extractable Nuclear Antigens (ENA) and Myositis Immunoblot, were accessed using the Nottingham University Hospitals Trust pathology database. Existing and accessible HRCT scans (n = 69), were evaluated, by a radiologist with a special interest in connective tissue disease-ILD. The Scleroderma Lung Study scoring system was employed, evaluating ground glass opacity, fibrosis, bronchiectasis and honeycombing in three anatomical zones. A binary logistic regression model evaluated the role of autoantibodies in ILD diagnosis.