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