

# Real World MDT Diagnosis of Idiopathic Pulmonary Fibrosis

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## INTRODUCTION

- Idiopathic pulmonary fibrosis (IPF) is the commonest idiopathic interstitial lung disease (ILD) and carries the worst prognosis, thus early diagnosis is key for initiation of disease-specific treatment.
- 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<sup>1</sup> though biopsy can afford significant morbidity and mortality.
- We aimed to evaluate our practice in diagnosing IPF in relation to these guidelines.

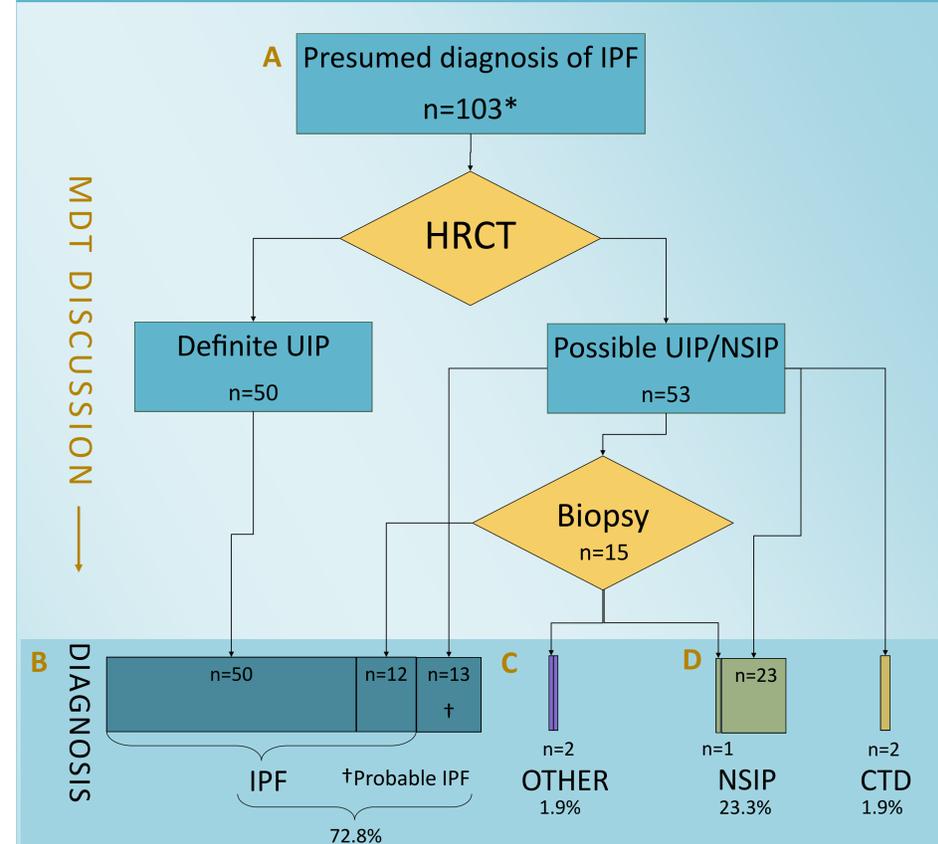
## METHODS

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

## RESULTS

- **Demographics:** 72 (70%) were male and mean  $\pm$  standard deviation age was  $69 \pm 8.2$  years.
- Only 49% patients had definite UIP pattern on HRCT. 51% had possible UIP or fibrotic non-specific interstitial pneumonitis (NSIP) based on ATS/ERS criteria.
- Of the fifty-three patients with possible UIP, almost one third (30%) patients had a lung biopsy, which confirms a UIP pattern in the majority of cases (87%).
- One patient died and one patient suffered with chronic pain post biopsy (13% of those biopsied).
- In the remaining two thirds of patients (70%), biopsy was not possible or not indicated (please see fig 2).
- Of the fifty-three patients with a radiological diagnosis of possible UIP/NSIP, thirteen patients were deemed to

**Figure 1 :** How diagnosis was made based upon MDT discussion, HRCT and biopsy



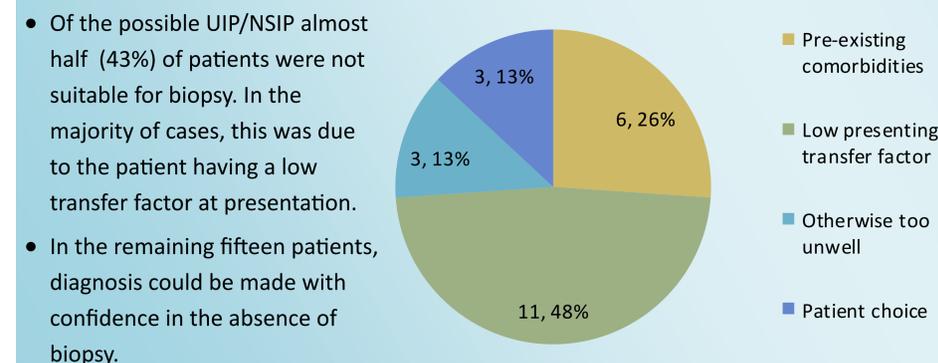
Decisions throughout the process of making the diagnosis were based upon HRCT, biopsy results and the clinical picture (disease progression, response to immunosuppressive therapy).  
 A: \*Originally n=104 though one patient discounted due to insufficient information regarding diagnostic process.

B: Diagnosis made. Length of bars proportionate to number of diagnoses.

C: Other diagnoses: DIP n=1; HSP n=1.

HRCT= High resolution computed tomography; IPF = Idiopathic pulmonary fibrosis; UIP= Usual interstitial pneumonitis pattern; NSIP= Non-specific interstitial pneumonitis; CTD= Connective tissue disease; HSP= Henoch-Schönlein Purpura (pulmonary involvement).

**Figure 2 :** Why were patients with possible UIP/NSIP not suitable for biopsy?



- Of the possible UIP/NSIP almost half (43%) of patients were not suitable for biopsy. In the majority of cases, this was due to the patient having a low transfer factor at presentation.
- In the remaining fifteen patients, diagnosis could be made with confidence in the absence of biopsy.

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 (26%) patients were clinically diagnosed as NSIP based on response to immunosuppression and stability of lung function.

## CONCLUSION

- 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.
- Lung biopsy is not without risk.
- Our clinical experience demonstrates that where there is diagnostic uncertainty, multi-disciplinary diagnosis of IPF increasingly utilises clinical data regarding progression and failed responses to immunosuppressive therapies.
- Considering the high yield of UIP after biopsy in patients with a radiological diagnosis of possible UIP/NSIP and concerns regarding morbidity and mortality, our practice has altered.
- In the subsequent year, 8.7% of patients underwent biopsy versus 14.6%, reflecting a change in practice.

## REFERENCES

1. *BTS Interstitial Lung Disease Guideline*; <https://www.brit-thoracic.org.uk/document-library/clinical-information/interstitial-lung-disease/ild-guidelines/bts-interstitial-lung-disease-guideline/>
2. Clinical Predictors of a Diagnosis of Idiopathic Pulmonary Fibrosis; CD Fell, FJ Martinez, LX et al; *Am J Respir Crit Care Med.* 2010 April 15; 181(8): 832–837. Published online 2010 January 7. doi: 10.1164/rccm.200906-0959OC