Irish Thoracic Society

Irish Thoracic Society **Interstitial Lung Disease Registry Annual Report 2018**

2018

Introduction

The first Annual Report from the ITS ILD Registry provides preliminary data on trends in the diagnosis, management and treatment of IPF in Ireland. The Registry will enable us to monitor standards of patient care and benchmark these against optimal diagnostic and care pathways. These are outlined in the ITS IPF Position Statement, launched in August 2018, which reflects our improved understanding of the pathogenesis of the disease and the introduction of new therapeutic options.

The main objective of the ITS ILD Registry is to advance our knowledge of ILD in Ireland by collecting and analysing patient information. The registry is designed to provide a record of ILD in Ireland, but also to provide clinicians with up-to-date information on how the disease is diagnosed and treated. It is our aim to provide a truly national registry so that all patients can participate no matter where they live or are diagnosed. The registry will also support research and aid in planning for future care resources in line with optimal pathways of diagnosis and care for people with ILD and IPF. The registry is not a prevalence study but will provide some estimates of disease frequency. At present we estimate that 1-2 new people receive a diagnosis of IPF each day in Ireland.

Methodology and Governance

The model for the ITS ILD Registry is identical to that used and approved in many hospitals in Ireland for cystic fibrosis. Patients give written informed consent at the hospital site and the consent record is maintained at that site. After informed consent, basic patient data relating to ILD is recorded in the registry. Data is pseudonymized in the registry with no identifiers related to clinical care. At each hospital site, the site principal investigator can view clinical and longitudinal information for each patient under their care.

All our systems and processes are in line with the highest standards of data protection and we are monitoring and reviewing these on an ongoing basis in line with evolving regulatory compliance requirements including GDPR. All data will be treated as confidential. No identifiable data will be issued to third parties. No participant will be identified in any presentations or publications of data arising from analysis of the registry.

Participating Centres

Cork University Hospital

Galway University Hospital

University Hospital Limerick

St Vincents University Hospital, Dublin

Tallaght Hospital, Dublin

St James's Hospital, The Mater Misericordiae University Hospital and Beaumont Hospital Dublin are expected to join in early 2019.

ITS ILD Registry Steering Committee

Chair: Prof. A O'Regan, Galway University Hospital, Galway

Vice-Chair: Dr. Michael Henry, Cork University Hospital, Wilton, Cork

Secretary: Ms. Suzanne McCormack, CEO, Irish Thoracic Society

CNS Rep: Ms Bernadette Bowen, Cork University Hospital Cork

CNS Rep: Ms Lindsay Brown, St Vincent's University Hospital Dublin

External: Dr. Colin Edwards, CEO, Merlin Consulting Ltd., Dublin

Member: Ms. Carita Bramhill, St. Vincent's University Hospital, Dublin

Member: Ms. Nicola Cassidy, Irish Lung Fibrosis Association, Dublin

ILD Nurse: Ms. Bridget Mulholland

Key Findings

We report data on the first 154 patients (there are approximately 200 in the registry but due to analysis we can only provide data on 154 this year).

The gender split is 68% male and 32% female. This compares to UK data where the gender split is 76% male to 24% female. The reason for higher proportion of women in the registry in Ireland is unclear.

19% of cases report a family history of IPF. This is also higher than UK figures which show that only 7% of cases report a family history.

Data on FVC and DLCO results indicates that approximately 75% of patients are being diagnosed at a stage in their disease progression where treatment can be administered. It also shows that 40% of patients have advance disease and could be candidates for lung transplant assessment. Approximately 80% of patients in the registry are receiving anti-fibrotic treatment.

Only 57% of cases completed a 6 Minute Walk Test (6MWT). This is significantly less than the figure reported in the British Thoracic Society ILD Registry Report (83%). 70% of patients are not on Oxygen therapy and only 38.6% were referred to Pulmonary Rehabilitation. These data suggest that all patients do not currently have access to a specialist physiotherapist who can provide exercise testing, pulmonary rehabilitation and oxygen assessment as prescribed in the ITS IPF Position Statement – there is a clear need to expand current services to cater for patients with IPF.

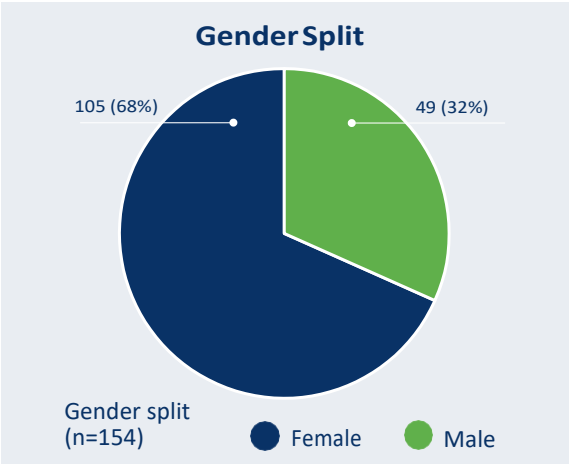
In terms of referral pathway - less than half of patients (46%) are coming through Primary Care – this indicates that increased awareness and education on the signs and symptoms of IPF is required at primary care level.

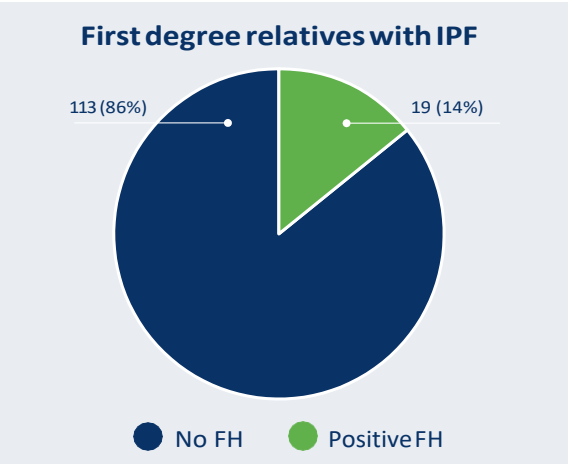
Although the data shows that 82.4% of cases have been discussed at MDT, this still falls short of the ITS position statement recommendation that all patients with possible IPF should receive an MDT based diagnosis and care pathway.

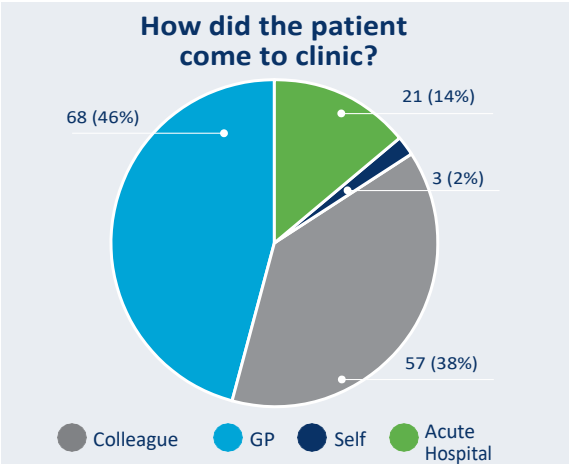
Ultimately most patients with IPF will progress to need for transplant or palliative care. In the registry 12% of patients have been referred for Lung transplant assessment and 13% to palliative care. Both figures suggest there is scope for improving access to these services. Particularly in the case of Lung Transplant which is recognised as a highly effective treatment for IPF. The ITS IPF Position Statement recommends that all patients with a diagnosis of IPF should be considered for transplant referral with an emphasis on prioritising patients with rapidly declining lung function, diffusion capacity (DLCO) below 40%, or respiratory failure.

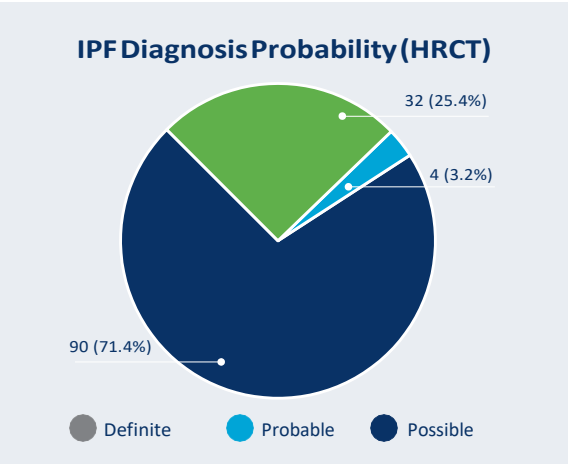
Total IPF Cases	Mean age at presentation
154	72.44

Smoking Status		
Status	**Number**	**%**
Current	27	18%
Former	89	58%
Never	36	23%
Not known	2	1%

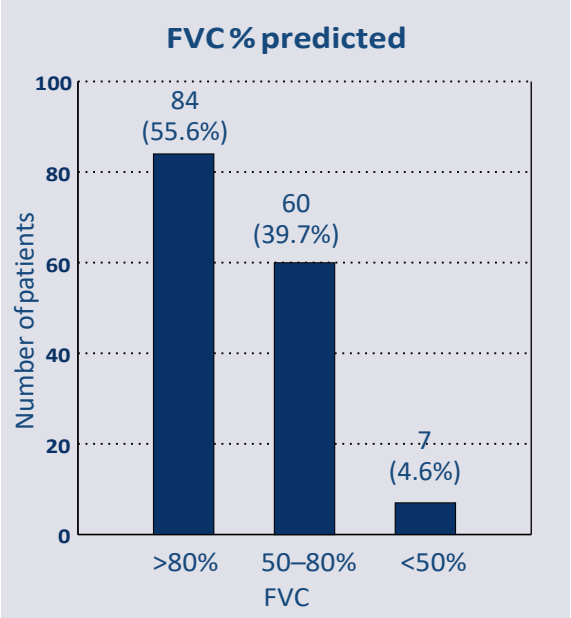


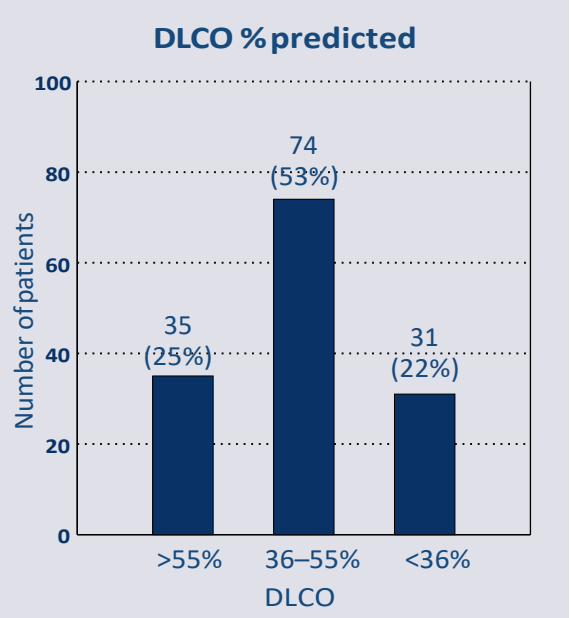






Symptom duration prior to presentation		
Duration	**Months**	**%**
Less than 6 months	55	42.3%
6 – 12 months	31	23.8%
12 – 24 months	24	18.5%
More than 24 months	20	15.4%





	Biopsy Number	%
Surgical	12	7.9%
Bronchoscopic	11	7.2%
Method of biopsy not known	10	6.5%
Not biopsied	120	78.4%
Total	153	100%

Histopathological confirmation of UIP pattern		
Biopsy method	High confidence	Low confidence
Bronchoscopic	7	2
Surgical	7	1
Degree of confidence not mentioned in 16 patients		

MDT Discussion	Number	%
Yes	126	82.4%
No	20	13.1%
Not known	7	4.5%
Grand total	153	100%

6MWT		
	Number	%
Yes	90	57%
No	63	39.9%
Not Known	5	3.1%
Grand total	158	100%

6MWT mean distance
328. +/– 112 metres

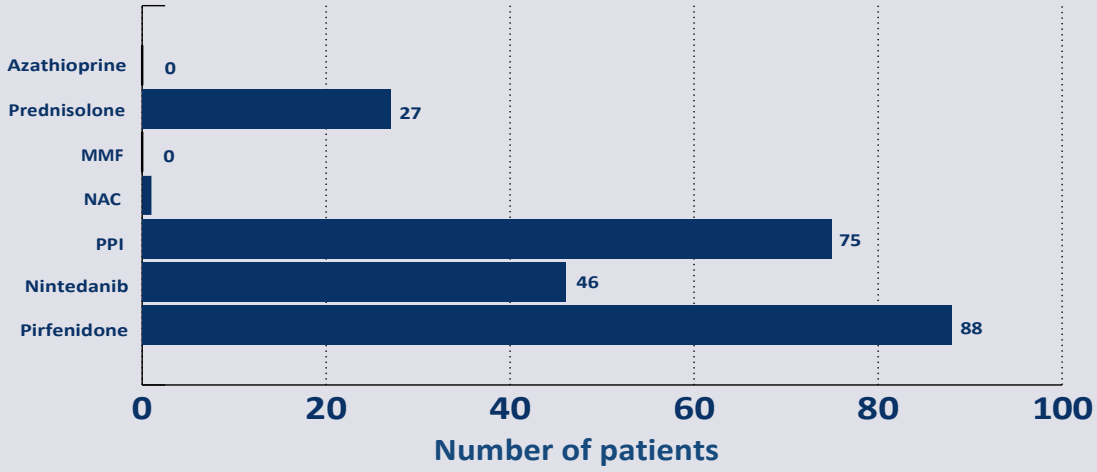
Is the patient on oxygen ?		
Oxygen	Number	%
Yes : LTOT	45	28.5%
Yes : Ambulatory	50	31.7%
No	110	70%
Grand Total	N/A	N/A

Pulmonary rehabilitation		
Patient referred	Number	%
Yes	61	38.6%
No	90	57.0%
Not known	7	4.4%
Patient declined	0	0.0%
Grand total	158	100%

Palliative Care		
Patient referred	Number	%
Yes	20	12.7%
No	114	72.2%
Not known	24	15.1%
Patient declined	0	0.0%
Grand total	158	100%

Autoimmune screen					
Autoimmune screen performed (n = 153)				Result	
	Yes	No	Not known	Positive	Negative
Rh Factor	94	23	36	10	84
Anti CCP	68	83	2	4	64
ANA	96	56	1	0	96
CPK	51	57	45	1	50
Extended myositis panel	0	11	142	0	0
ENA	74	45	34	3	71

Current medications



Antifibrotic treatment		
	Number	%
Pirfenidone	88	57.5%
Nintedanib	46	30.1%
Not known	18	11.8%
Grand total	153	100%

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For further information go to:
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