



Irish Thoracic Society

Irish Thoracic Society **Position Statement** **on the Management** **of Idiopathic** **Pulmonary Fibrosis**

The Irish Thoracic Society
Interstitial Lung Disease Group
AUGUST 2018

CONTENTS

1. **Introduction**
 2. **Epidemiology**
 3. **Optimal pathways for the delivery of care to patients with IPF**
 - a. Education and Awareness
 - b. The National Patient Charter for IPF
 - c. Diagnosis
 - d. Multi-disciplinary Clinical Care
 - e. National Programmes - access to treatment and advocacy
 4. **Conclusion**
- Appendices**

Members of the ITS ILD Group:

Chair: Professor Anthony O'Regan
Consultant Respiratory Physician, Galway University Hospital, Galway

Ms Lindsay Brown
Respiratory Advanced Nurse Practitioner, St Vincent's University Hospital, Dublin

Dr John Bruzzi
Consultant Radiologist, Galway University Hospital, Galway

Ms Nicola Cassidy
Director, Irish Lung Fibrosis Association (ILFA)

Professor Seamas Donnelly
Consultant Respiratory Physician, Tallaght Hospital, Dublin

Professor Jim Egan
Consultant Respiratory Physician, Mater Misericordiae University Hospital, Dublin

Dr Aurelie Fabre
Consultant Histopathologist, St Vincent's University Hospital Dublin

Dr Ruairi Fahy
Consultant Respiratory Physician, St James's Hospital, Dublin

Ms Tara Hannon
Senior Physiotherapist, Galway University Hospital, Galway

Dr Michael Henry
Consultant Respiratory Physician, Cork University Hospital, Cork

Dr Killian Hurley
Consultant Respiratory Physician, Beaumont Hospital, Dublin

Professor Michael Keane
Consultant Respiratory Physician, St Vincent's University Hospital, Dublin

Ms Suzanne McCormack
CEO, Irish Thoracic Society

Dr Emmet McGrath
Consultant Respiratory Physician, St Vincent's University Hospital, Dublin

Dr Aidan O'Brien
Consultant Respiratory Physician, Limerick University Hospital, Limerick

Dr Kate O'Reilly
Consultant Respiratory Physician, Mater Misericordiae University Hospital, Dublin

Professor Karen Redmond
Consultant Thoracic & Lung Transplant Surgeon, Mater Misericordiae University Hospital, Dublin

Acknowledgements

With thanks to Mr Matt Cullen for his review of the Position Statement on the Management of Idiopathic Pulmonary Fibrosis from a patient's perspective.

Thanks also to Ms Bettina Korn, End-of Life Care Co-ordinator, Hospice Friendly Hospitals Programme, St James's Hospital Dublin, for review of and contribution to the Palliative Care section.

Thanks to Dr Michael Dillon for his assistance in the early drafting of this statement.

Thanks to the Irish Lung Fibrosis Association (ILFA) for kindly supporting the design and print of the ITS Position Statement on the Management of Idiopathic Pulmonary Fibrosis.

1

INTRODUCTION

Idiopathic Pulmonary Fibrosis (IPF) is a chronic, progressive and fatal lung disease resulting in severe morbidity with shortness of breath and impaired quality of life due to worsening respiratory function. The Irish Thoracic Society (ITS) Interstitial Lung Disease (ILD) Group is committed to supporting the development and delivery of best standards of care to patients with IPF and related ILDs.

The last ITS IPF Position Statement was published in 2012. Since then there have been major changes in diagnostic pathways, treatment options and care plans for patients with IPF and other related ILDs. The updated ITS IPF Position Statement incorporates the advances which have occurred since 2012 and provides guidance to healthcare providers on the optimal diagnostic and care pathways.

The ITS Position Statement on the Management of IPF is also aimed at policy and decision makers as it outlines the resources needed to provide fair and equitable access to the standards of care that our patients with IPF and related ILDs need and deserve.

2

EPIDEMIOLOGY

IPF is the most common type of ILD. While it is sometimes described as rare, it occurs with a similar frequency to that of stomach, brain, and testicular cancers and its prognosis is worse than many other cancers. A review of global incidence and mortality data has suggested a conservative incidence range of 9 cases per 100,000 per year in Europe. However, recent data from the UK suggests that there is a significant under-estimation of the disease prevalence. The British Lung Foundation recently reported an incidence of 6000 new cases of IPF, with 5300 people dying of this disease in the UK each year. Like the UK, it is probable that this condition is more common than predicted in Ireland and would extrapolate to over 400 new cases per year and a prevalence of over 1000 cases. This may reflect environmental and genetic risk factors but more research is required in this area. IPF is more common in men and rare in people under the age of 50 years. International studies demonstrate a median survival time of 3 years from diagnosis, however the prognosis is variable with some patients experiencing longer survival times dependent on disease stage. It is anticipated that recent advances in treatments will also improve both the quality and duration of survival. There are limited data on the disease characteristics and incidence in Ireland.

In order to improve knowledge of IPF in Ireland, the ITS ILD Group has developed a National Patient Registry to record the incidence of and clinical data on patients with IPF. This spans all the hospital groups with most specialist centres actively participating. Expansion of the registry is clearly desirable so that maximum patient inclusion is reached and patient reported outcome measures (PROMs) can be captured and evaluated. While broad-based funding is welcome, the ITS ILD Group recommends that primary funding for the registry should be provided by the Department of Health (as is the case for the National Cancer Registry) or the Health Service Executive (HSE). The establishment of an adequately funded independent register with an appointed national co-ordinator and data manager would provide much needed accurate data on the incidence, prevalence and patient outcomes and would be of significant pharmaco-economic benefit in planning for future care resources. The ITS ILD Registry has a vision to engage with other international ILD Registries and Thoracic Societies.

3

OPTIMAL PATHWAYS AND CLINICAL STRUCTURES FOR THE DELIVERY OF CARE TO PATIENTS WITH IPF

a. Education and Awareness

As there are currently two effective anti-fibrotic disease-modifying drugs approved for use in Ireland (Pirfenidone and Nintedanib), the importance of an early and accurate diagnosis and rapid referral to centres with expertise in IPF cannot be overstated. Early diagnosis and rapid referral will require the development of effective awareness campaigns to educate the public, general practitioners and integrated care clinicians of the signs and symptoms of the disease. Education programs with the Irish College of General Practitioners and radiology professional bodies should be developed to ensure early recognition of the clinical features of IPF.

b. The National Patient Charter for IPF

Once a diagnosis of IPF is suspected the ITSILD Group recognises the need to deliver the key areas of The National Patient Charter for IPF developed by the Irish Lung Fibrosis Association (ILFA) in 2015 (see below). Patients should be partners in developing their individual care plan and should be informed of the full range of treatment options available to them including pharmacological, non-pharmacological, surgical and palliative care interventions.

The National Patient Charter for IPF

This charter informs patients of the six key areas of care that they should be entitled to:

- 1) Early and accurate diagnosis
- 2) Clear information about IPF
- 3) Access to medication and oxygen
- 4) Access to pulmonary rehabilitation and exercise programmes
- 5) Early referral to the national lung transplant unit with a minimal emphasis on age
- 6) Access to psychological and palliative care services.

c. Diagnosis

Multi-Disciplinary Teams (MDTs) working at ILDSpecialist Centres (see Appendix 2) are required to develop the clinical structures required to facilitate an early, accurate and confident diagnosis and deliver optimal care to patients with IPF. Given geographic challenges, the ITS supports the establishment of specific centres with expertise in ILDS and IPF. These centres must be resourced and structured so that there is fair and equitable access to the expertise in these centres for all patients no matter where they reside or where they receive their initial diagnosis. It is recommended that patients referred with suspected IPF should expect to be reviewed with the diagnosis confirmed and treatment initiated within 16 weeks. This will require access to healthcare professionals with expertise in ILDS and to high resolution CT scanning, pulmonary function testing and biopsy when required. The need for tissue diagnosis should be evaluated by an MDT discussion. Biopsies (transbronchial, cryobiopsies, Video Assisted Thoracoscopic Surgery (VATS) / surgical) should be specifically targeted and the mode specified by the MDT. Currently the resources provided for this condition are insufficient to reliably meet this guideline.

Each patient with possible IPF should receive an accurate diagnosis by a recognised MDT-based pathway in a specialist centre (see Appendix 2) with the following specialists:

- 1 Respiratory Consultant with special interest in ILD
- 2 Radiologist with special interest in ILD
- 3 Access to a Histopathologist with special interest in ILD
- 4 Access to Cardiothoracic Surgery
- 5 Access to Advanced Thoracic Bronchoscopy
- 6 Access to Accredited Pulmonary Function Laboratory

d. Multi-disciplinary Clinical Care

Once the diagnosis is confirmed, patients should be linked to an MDT for ongoing treatment. There is a need for rapid and accessible clinical support to answer questions and address concerns in a timely manner. The ITS ILD Group supports a 'hub and spoke' model of shared care. There should be adequately resourced specialist centres in each hospital group providing diagnosis and support pathways to address acute and chronic ongoing issues such as medication side-effects, transplant referrals and acute exacerbations. The management of this disease requires a combination of education, physiological and pulmonary rehabilitation assessments, appropriate pharmacological and non-pharmacological care, access to lung transplant assessments and palliative care. Psychological support is required to assist with the impact of this diagnosis on patients and their families.

A specialised multi-disciplinary clinical team delivering ongoing treatment should include:

- 1 A respiratory consultant with experience in ILD and with access to a respiratory consultant with a special interest in ILD.
- 2 ILD Specialist Nurse (circa 1 per 50 patients).
- 3 Specialist Respiratory Physiotherapist with an interest in ILD.
- 4 There should be access to thoracic surgery, rheumatology, palliative care, as well as medical social worker, psychological and dietetic supports.

Staffing levels should be sufficient to allow cross-cover during annual leave.

The clinic should be dedicated to ILD and provide adequate access to Pulmonary Function Tests, Six-Minute Walk Tests, symptom review, treatment review including oxygen therapy, and review of treatment related side-effects including liver function and other laboratory testing. The expertise to provide timely discussion around and referral to transplant services should be available.

Current anti-fibrotic treatments for IPF have notable side-effects which may require considerable face-to-face time between the patient, the physician and the respiratory nurse specialist to ensure safe use of the medicines. Ongoing multiple interactions between patients and healthcare professionals are needed to maximise efficacious safe treatment and to support patients' psychological adjustment to living with IPF. An individualised care plan that takes the progressive and uncertain nature of IPF into account should be developed in collaboration with all patients.

Education should be delivered by skilled clinicians to allow a clear understanding of the condition, its prognosis and the treatment options. A patient-centred approach encompassing pharmacological, non-pharmacological and holistic treatments is needed. Once diagnosed, patients should be informed of the information and support available from the Irish Lung Fibrosis Association (ILFA) (www.ilfa.ie), the national patient organisation for IPF.

Structured systems need to be established to ensure that patients have access to essential services such as pulmonary rehabilitation, nutritional advice, medical social work input, psychological and palliative supports and referral to lung transplantation services in a timely and seamless fashion.

i. Clinical Nurse Specialists

Clinical Nurse Specialists are critical to the delivery of patient centred care as part of the multi-disciplinary team. All clinics should be supported by a Clinical Nurse Specialist with specific training in ILD (and where possible Advanced Nurse Practitioner) to provide nurse-led clinics and structured telephone follow-ups. IPF nurse-led clinics have been shown to be an effective way of dealing with patient enquiries and management of treatment side-effects.

ii. Physiotherapy

All patients should have access to a specialist physiotherapist who can provide the following:

- 1** Exercise testing (e.g. six minute walk test) with review of current functional status;
- 2** Bespoke Pulmonary Rehabilitation Programme supported by a structured home exercise programme and education on pacing and energy conservation;
- 3** Monitor disease progression and provide support and symptom management techniques, aids and appliances;
- 4** Oxygen assessment – both ambulatory and continuous;
- 5** Treatment and education for chronic cough and dysfunctional breathing.

At present the majority of Pulmonary Rehabilitation services target patients with Chronic Obstructive Pulmonary Disease (COPD). There is a clear need to expand resources to cater for patients with IPF and to ensure adequate management of long-term oxygen therapy.

iii. Lung Transplant

Lung transplant is recognised as a highly effective treatment for IPF and in Ireland over 30% of patients receiving lung transplantation have IPF. Appropriate and timely referral to the National Lung Transplant Centre at the Mater University Hospital is required. All patients should be considered for transplant referral with clear emphasis on prioritising patients with rapidly declining lung function, diffusion capacity (DLCO) below 40%, or respiratory failure. In some highly selected cases, transplant can be considered over the age of 70 years. The group recognises the need for adequate resources in the National Transplant Centre and also the ability of each referring centre to actively participate in pre-transplant assessment by working closely with the National Transplant Centre. There should be a targeted proforma linked to an electronic referral process to improve communication and collaboration. To optimise care, the transplant service needs to be supported to perform single lung transplants simultaneously.

iv. Palliative Care

There should be timely and adequate access to community, general and specialist palliative care services. The major aims of palliative care in IPF are to improve quality of life by addressing symptom-related, psychological, social and spiritual needs, and to ensure that patients with IPF live well throughout the course of the disease. Clinicians in ILD specialist centres and those managing ongoing care should adopt a person-centred approach to care that is responsive to patients' palliative care needs as and when they arise throughout the disease trajectory. In view of the life-limiting nature of IPF, the assessment for and the management of patients' palliative care needs should be a core skill of all clinicians caring for patients with IPF. This includes open and sensitive communication about prognosis, the goal of treatment, end-of-life care options and advance care planning. ILD services should develop formal links with Specialist Palliative Care Services in order to refer patients appropriately (e.g. for breathlessness management clinics, community services etc.), develop shared care protocols and enhance inter-disciplinary knowledge transfer in order to improve patients' quality of life.

e. National Programmes to ensure optimal access to treatment and advocacy

At a national level consideration should be given to the establishment of a National Clinical Programme for IPF, as has been done for other respiratory conditions such as Asthma, Chronic Obstructive Pulmonary Disease, Cystic Fibrosis, and 31 other disease areas.

The National Centre for Pharmacoeconomics should engage with Specialist ILD Physicians and with ILFA during the evaluation process of new evidence-based IPF medications and therapies, to ensure that clinician and patient perspectives are included. As further progress in the pharmacological treatment of IPF is expected, it is important that IPF patients in Ireland have access to investigational drugs by participation in clinical trials. A list of centres interested in IPF clinical trials is available from the ITS.

Screening programmes for familial IPF may be possible in the future and there is an evolving role for genetic testing in IPF. The need for genetic counselling and the legal and indemnity implications for individuals must be considered. There is a vision that Ireland will actively engage and lead in the area of IPF research, including healthcare related outcomes, genetic susceptibility, and other areas of pathogenesis.

The complex and multi-disciplinary nature of an ideal IPF Care Pathway is shown in Appendix 1.

CONCLUSION

For many years, ILDs and IPF have been something of a 'Cinderella' of respiratory diseases most probably because of the absence of effective treatments. However, this has now changed and it is the view of the ITS that the standards of diagnosis and management of IPF need to be radically improved as outlined above. All patients with IPF should have access to specialist centres and all appropriate treatments without undue delay.

References available on request.

IPF MULTIDISCIPLINARY CARE PATHWAY

Visit 1 Initial Medical Consultation

Investigations Scheduled: HRCT, Bloods, SpO2, PFTs

MDT Discussion



Visit 2 Results of Investigations:

- Definitive IPF diagnosis given
- Confirm and explain diagnosis
- Discuss implications
- Advise of treatment options
- Explain monitoring process
- CNS and HSCP complete functional and psychosocial assessments
- Provide literature
- Give time to think about anti fibrotic therapy - 1 month

Visit 2 or 3: Personalised Care Plan Devised

Patient Education	Anti-Fibrotic Therapy	Monitoring	Pulmonary Rehab	Oxygen Referral
Smoking cessation		Resting SpO2, RR	PCCC	
Nutritional advice		Baseline 6MWT	Dietician	
Physio referral		Spirometry +DLCO	OT	
Vaccinations		LFTs	MSW	
ILFA		Symptom Scores	PHN	
		Weight	Psychologist	
		Symptom Management		
		Exacerbations/hospitalisations		

Visit 4: 6 weeks from commencing Anti-fibrotic Rx.

- Revise Care Plan
- Drug Tolerance
- Dose adjustment
- Monitoring: PFTs, RR, SpO2, LFTs, 6MWTs
- Symptom Management
- Activate Support Services
- Correct exertional desaturation

Visit 5: onwards

- 3-6 monthly review depending upon signs of disease progression.
- Revise Care Plan

Respiratory CNS

Telephone Advice Line

Earlier Review if appropriate

Clinical Trial Enrolment

Exacerbation Admission

Transplant Referral
DLCO < 40% predicted
FVC declines by 10%

Palliative Care
Think Ahead
Advanced Directive
Breathlessness Clinics
End of Life Care

Glossary

HRCT – High Resolution CT Scan
SpO2 – Oxygen Saturation
PFT – Pulmonary Function Test
MDT – Multidisciplinary Team

CNS – Clinical Nurse Specialist
HSCP – Health and Social Care Professional
ILFA – Irish Lung Fibrosis Association

RR – Respiratory Rate
6MWT – 6 Minute Walk Test
DLCO – Diffusion Capacity
LFT – Liver Function Test
PCCC – Primary Care and

Community Care
OT – Occupational Therapy
MSW – Medical Social Work
PHN – Public Health Nurse
FVC – Forced Vital Capacity

Regional Specialist ILD Centres Contact List
Beaumont Hospital, RCSI Hospital Group

Lead Physician: Dr Killian Hurley
Email: killianhurley@beaumont.ie

Secretary: Ms Joy Bennett
Tel: 01 809 3058 *Fax:* 01 797 4714

Specialist Nurse:
 Ms Catherine McGeoghegan
Email: respnurses@beaumont.ie
Tel: 01 809 2384

Cork University Hospital

Lead Physician: Dr Michael Henry
Email: michael.henry@hse.ie

Secretary: Ms Noreen Finnegan
Tel: 021 4920169 *Fax:* 021 4920168

Specialist Nurse: Ms Bernadette Bowen
Email: bernadette.bowen@hse.ie
Tel: 086 8228176

Galway University Hospital

Lead physician: Professor Anthony O Regan

Secretary: Ms Cora Hallinan
Email: cora.hallinan@hse.ie
Tel: 091 755225

Specialist Nurse: Ms Donna Langan
Email: donna.langan@hse.ie
Tel: 091 544126

Specialist Nurse: Ms Helen Mulryan
Email: helen.mulryan@hse.ie
Tel: 091 544126

Senior Respiratory Physiotherapist:
 Ms Tara Hannon
Email: tara.hannon@hse.ie
Tel: 091 544126

Mater Misericordiae University Hospital

Lead Physician: Dr Kate O'Reilly

Secretary: Ms Mairead Fewer
Tel: 01 803 4547 *Fax:* 01 803 4180

Specialist Nurse: Ms Lynn Fox
Tel: 01 803 2000 (switch) Bleep 4824

St James's Hospital Dublin

Lead Physician: Dr Ruairi J Fahy MD, FCCP
Email: rfahy@stjames.ie

Secretary: Ms Ruth Kelly
Email: fahysec@stjames.ie

Tel: 01 416 2339 *Specialist*
Fax: 01 410 3765

Nurse: Ms Roisin Kennedy
Email: rukennedy@stjames.ie
Tel: 01 410 3763

St Vincent's University Hospital

Lead Physicians
 Professor Michael P Keane
Email: michael.p.keane@ucd.ie

Dr Emmet McGrath
Email: emmetmcgrath@svhg.ie

Admin Manager Respiratory:
 Ms Georgina O'Reilly
Email: g.oreilly@st_vincent's.ie

Specialist Nurses:
 Ms Lindsay Brown
Email: lindsay.brown@svuh.ie
Tel 01 221 4000 *Pager* 771

Ms Patricia Jones
Email: p.jones@svuh.ie
Tel 01 221 4000 *Pager* 168
Fax 01 221 3708

Tallaght University Hospital & Trinity College Dublin

Lead Physician:
 Professor Seamas Donnelly

Pulmonary Fibrosis Referral Centre, Medicine, Trinity Centre, Tallaght University Hospital, Dublin 24

ILD Clinical Appointments Secretary:
 Ms Lorraine Flood
Email: lorraine.flood@amnch.ie
Tel: 01 414 4608 *Fax:* 01 896 2988

Specialist Nurse: Ms Mary Poland
Email: mary.poland@amnch.ie
Tel: 01 414 4608

University Hospital Limerick

Lead Physicians:
 Dr Aidan O'Brien
 Dr Brian Casserly

Secretary: Ms Mairead O'Brien
Tel: 061 585661 *Fax:* 061 482921

Specialist Nurses:

Ms Paula Ryan
Email: paula.ryan@hse.ie

Ms Maria Cullinan
Email: maria.cullinan@hse.ie

Ms Noelle Ryan
Email: noelleb.ryan@hse.ie

Ms Eilis Gleeson
Email: eilis.gleeson@hse.ie



Irish Thoracic Society

For further information contact:

The Irish Thoracic Society
E: info@irishthoracicsociety.com
T: 01 5677201
www.irishthoracicsociety.com

For patient information and support contact:

The Irish Lung Fibrosis Association
E: info@ilfa.ie
T: 086 871 5264
www.ilfa.ie

Irish Thoracic Society
Creidim Centre, Leopardstown Rd
Dublin 18

T: +353 (0) 1 567 7201
E: info@irishthoracicsociety.com
W: www.irishthoracicsociety.com