BTS ILD Registry Annual Report 2021: a summary of the UK IPF Registry for the general public

Idiopathic pulmonary fibrosis (IPF) is an incurable lung disease in which scars are formed in the lung tissues. It is a devastating condition characterised by increasing breathlessness, cough and fatigue. The symptoms may also impact on mental health causing problems such as anxiety and depression.

Life expectancy for people living with IPF remains poor, with only a quarter of people surviving more than five years from diagnosis. IPF has a worse life expectancy than most cancers and information collected on the UK IPF Registry has shown that there are still significant delays from initial symptoms to diagnosis.

It is estimated that around 30,000 people in the UK live with IPF, yet there are limited treatment options available. Over 5,000 people in the UK die every year from the disease which accounts for 1% of all deaths.

While the treatment options for IPF remain limited, significant advances have been made in the care of patients with IPF over recent years. These include the introduction of antifibrotic medications aiming to slow the progression of the disease and publication by the National Institute for Health & Care Excellence (NICE) of the UK Quality Standard for the management of IPF.

Why is a registry needed?

Patient registries are collections of healthcare data on people living with a particular disease or condition. They aim to improve outcomes by making it possible to track treatment and care and link this to clinical outcomes of people living with the disease.

It is only by collecting information about people living with IPF that researchers, people treating patients, and people making decisions about healthcare policies can properly understand the impact these advances have on people living with IPF.

The British Thoracic Society (BTS) set up the BTS Interstitial Lung Disease (ILD) Registry in 2013. It covers two diseases: IPF and sarcoidosis. Information is submitted by hospitals on behalf of the patients they treat. The aim is to include as many people as possible in the Registry. BTS encourages hospitals and patients to agree to their data being collected so that the information in the Registry is as rich as possible.

Who is on the Registry?

The UK IPF Registry is one of the largest and most comprehensive in the world. Any patient with definite or strongly suspected IPF diagnosed from 2013 onwards can be included so long as they give their consent. As of 30th June 2021, a total of 3,385 patients have been registered (an increase of 588 patients since the end of June 2020). A total of 75 centres across the UK have so far been approved to participate in the Registry.

What information is collected for the Registry?

The BTS ILD Registry now includes nearly nine years of data, which makes it possible to analyse trends and helps us to gain a greater understanding of the disease.

The Registry includes **demographic data**, including the patient's gender, age and other medical problems in addition to IPF. From the data collected we know that:

- 79% of patients are male and this has remained constant over the lifetime of the Registry
- People with IPF are on average 74 years old when first seen in hospital and 72% of patients are aged 70 and over.
- 84% of patients with IPF also have other medical problems. These have remained stable over the lifetime of the Registry, with the most common being high blood pressure (42%), ischaemic heart disease (28%), diabetes (24%) and gastro-oesophageal reflux disease (24%).

The Registry also includes **clinical information** such as how long patients had symptoms prior to diagnosis and how long patients had to wait from referral to their first hospital clinic visit. For example, from the data collected since 2013, we know that unfortunately patients have symptoms for a considerable period prior to diagnosis, and that on average 64% of patients have had symptoms for more than 12 months before their first hospital clinic visit. We also know that on average the time a patient waits from referral to the first hospital clinic visit is 13.5 weeks. This highlights that there is still work to be done in raising awareness of IPF and getting people assessed more quickly. By continuing to collect this information we can understand whether the NHS nationally is meeting the NICE Quality Standard and whether there are specific areas in which care for patients with IPF can be improved.

The Registry also collects information on how patients are diagnosed. For example, we know most patients are

diagnosed using HRCT (high-resolution computed tomography) scanning. We also know that most cases are discussed at a specialist multidisciplinary team (MDT) meeting with 92% of cases being discussed at the first clinic visit. Only a very small proportion of patients (currently just under 3%) require a lung biopsy to confirm the diagnosis and the use of lung biopsies has declined over the past 8 years, especially since the diagnostic guidelines were updated in 2018.

The Registry also captures important information on the proportion of patients participating in clinical trials. For example, we know that over the lifetime of the Registry the proportion of patients recruited to clinical trials has remained low. More work needs to be done to improve access to clinical trials for patients with IPF in the future.

Hospitals are able to monitor their performance using Registry data and access data on their own patients. A local 'dashboard' is generated by the Registry every six months, which allows hospital healthcare professionals to keep an eye on how their service is running. Information which identifies individual patients is only available to the submitting hospital (that is, to the team responsible for treating the individual patients).

BTS can analyse data for the whole country to help improve clinical care and when designing clinical trials or recruiting patients to trials. However, information which identifies individual patients cannot be seen in this national data.

Recent developments and plans for the future of the UK IPF Registry

It has been a challenging year for the medical community as the respiratory workforce, in particular, has continued to turn its attention to managing patients with COVID-19 throughout 2021. Perhaps unsurprisingly this is reflected in a fall in the number of entries to the UK IPF Registry in the first six months of 2021. There were only 64 new patient entries in the first half of 2021 compared to 245 entries in the first half of 2019. This may limit the interpretation of recent data, but we hope, with the continued effort from everyone involved, that the Registry will continue to grow from strength to strength as we recover from the COVID-19 pandemic.

It is exciting that the UK IPF Registry is now being used to collect data for use in a new randomised clinical trial called 'Treating idiopathic Pulmonary Fibrosis with the Addition of Lansoprazole (TIPAL)'. This study is being run by the Norwich Clinical Trials Unit and is financed by the National Institute of Health Research (NIHR). It involves directly capturing lung function and other relevant data from the BTS IPF registry for patients participating in the trial. This study is open for recruitment from all sites working with the registry. More information is available from tipal@uea.ac.uk . We hope that this collaboration will serve to strengthen the quality of data collected during this trial.

The future of the Registry depends on patients consenting to their information being used to help researchers, contribute to future planning of healthcare services and drive-up standards of care for all patients with IPF. We hope that BTS can continue to build on the success of the Registry for many years to come.

Information for the public

This document has been prepared by Mr Steve Jones and Dr Wendy Funston, on behalf of the BTS ILD Registry Steering Group, as a brief summary of the content and key points from the BTS ILD Registry Annual Report 2021. If you have any queries about the report and your personal medical circumstances please discuss these with your health care professional.

The charity Action for Pulmonary Fibrosis supports patients who have been diagnosed with pulmonary fibrosis https://www.actionpulmonaryfibrosis.org/

The full report is available on the BTS website at: https://www.brit-thoracic.org.uk/quality-improvement/lung-disease-registries/.

The content of this document may be used by health care professionals in discussions with patients and/or carers, but the source of the material must be acknowledged.

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