BTS ILD Registry Annual Report 2020: 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, disability and death, three to four years after diagnosis. Only 25% of people survive for five years. IPF has a worse life expectancy than most cancers. The symptoms also impact on mental health causing problems such as depression and anxiety.

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.

Although treatment options for IPF remain limited, there have been significant advances in the management of patients with IPF over recent years. These include the introduction of new antifibrotic drugs and the publication by the National Institute for Health and Care Excellence (NICE) of a new 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 2020, a total of 2,797 patients with IPF were included on the Registry (an increase of 323 patients (+13%) compared to October 2019). The UK IPF Registry continues to go from strength to strength. A total of 73 centres across the UK have so far been approved to participate in the Registry.

What information is collected for the Registry?

In 2020, a review of the entire UK IPF Registry dataset was conducted, which makes it possible to analyse data trends over the past 7 years.

The Registry includes demographic data, including the patient's gender, age and comorbidities. This helps us to understand who is affected by IPF. From the data collected over the past 7 years we know that:

- 78% of patients are male, and this has remained consistent over the lifetime of the Registry
- People with IPF are on average 73.6 years old when first seen by hospital, and that 71% of IPF patients are aged 70 and over.
- Many patients with IPF have other comorbidities, with the most common being high blood pressure (44%), ischaemic heart disease (27%), diabetes (25%) and gastro-oesophageal reflux disease (25%).

The Registry also includes clinical information such as how long it took for patients to be referred to the clinic, their lung function test results, and what treatments they are receiving. For example, from the data collected since 2013, we know that on average 41.5% of patients have reported chest symptoms for more than 2 years before they first attended a clinic. This highlights that there is still work to be done in raising awareness of IPF and getting people assessed more quickly. By collecting this clinical information, we can understand whether the NHS nationally is meeting the NICE Quality Standard and whether there are focused areas in which care for patients with IPF can be improved.

The Registry also includes information on how patients are diagnosed. For example, we know that most patients are diagnosed based on their symptoms and using high resolution computed tomography (HRCT) scanning. Only a very small proportion of patients require a lung biopsy to confirm the diagnosis. The use of lung biopsies has declined over the past 7 years especially since the update of the diagnostic guidelines in 2018.

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Important information on participation in clinical trials is also captured through the Registry. For example, we know that the proportion of patients recruited to clinical trials has remained consistently low throughout the lifetime of the UK IPF Registry. 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. The Registry generates a local 'dashboard' 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 (i.e. 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

There have been several developments over the last year since the 2019 BTS ILD Registry Annual Report was published. In early 2020 BTS opened a 'data access request process', which allows external organisations to apply to BTS to access some of the Registry data to carry out research. It aims to benefit future patients by generating new insights into the diagnosis and treatment of IPF.

Members of the BTS ILD Registry Steering Group have recently analysed Registry data to compare the different ways predicted lung function values are calculated. This is important as predicted lung function values are used to monitor disease progression and to determine whether patients with IPF are eligible for antifibrotic medication. The results of this research were presented at the 2020 European Respiratory Society Congress and highlighted the importance of standardising lung function calculations in order to ensure that all patients with who are eligible for antifibrotic medications have access to treatment.

Other recent developments for the Registry include supporting clinical trial involvement. For the first time, the UK IPF Registry is being used to collect data for use in a large clinical trial. Approval has been given for UK IPF Registry data to be used in the randomised trial 'Treating Idiopathic Pulmonary Fibrosis with the Addition of Lansoprazole' (TIPAL), where patients are involved in both the trial and the UK IPF Registry. We hope that this will serve to strengthen the quality of data collected during this trial. This study is funded by the National Institute of Health Research.

Perhaps unsurprisingly, there has been a fall in the number of new IPF patients registered in 2020, as the respiratory community has turned its attention to manage patients with COVID-19 infection. This may limit the interpretation of recent data, but we hope, with the continued effort from everyone involved, that the Registry can continue to go from strength to strength.

The future of the Registry is reliant 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 2020. 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.

British Thoracic Society
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