# BTS ILD Registry Annual Report 2021

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This work forms part of the BTS Respiratory Quality Improvement activities. We work with our members, healthcare professionals from other specialties, and patients and carers to improve standards of care for people with respiratory diseases, and to support those who provide that care.

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# FOREWORD: DATA SHARED TO IMPROVE CARE

This is the 6th annual report from the BTS Interstitial Lung Disease Registry, which includes the UK IPF Registry and the UK Sarcoidosis Registry. It is a database of over 4,100 consenting people with the most common forms of interstitial lung disease: idiopathic pulmonary fibrosis and sarcoidosis. The data were collected over the last eight years from 75 participating centres across the UK. As such, it is one of the largest interstitial lung disease registries in the world.

The purpose of the Registry is to improve care and outcomes for people with interstitial lung disease by collecting demographic, clinical and follow-up data – both prospectively and retrospectively – from people with idiopathic pulmonary fibrosis or sarcoidosis throughout the UK. Participating sites, which submit data voluntarily, are able to compare their real-time data and outcomes with the rest of the UK with the aim of ensuring best practice.

This link between data and best practice is key to the Registry.

With this is mind our objectives are that:

- **Clinicians** use the Registry as a working tool to support patient monitoring and service benchmarking;
- **Commissioners** and policy-makers use the Registry to understand the burden of disease and ensure interstitial lung disease services are adequately resourced; and
- **Academics** use the Registry to gain insight into these conditions so that patient outcomes can be improved.

BTS has been working alongside NHSE since for a number of years to update the NHSE ILD Quality Dashboard, designing items which will be easy for centres to capture and to provide a true representation of local delivery and standards of care. Full details of the updated NHSE ILD Quality Dashboard items are included on the BTS website.

The COVID-19 pandemic has altered all of our lives - none more than those of the medical profession. It has been a challenging year for medical community and in particular the respiratory workforce has been deployed to manage the increasing numbers of inpatient admissions with COVID-19. Referral patterns, clinical practice, workload and priorities have all changed. Data entry completeness has therefore, understandably, reduced. Furthermore, the timeframe of the report year has been modified so that 2021 data is are line with other BTS reports. All of these factors meant that comparison between 2021 data and data from previous years should be interpreted with caution.

The UK IPF Registry is open to all UK hospitals (once they have gained local Caldicott approval). Most of the participating centres are secondary care hospitals. However, the majority of patients on the Registry come from tertiary specialist centres. This has implications for the interpretation of some aspects of the Registry, including referral route, prescribing practices and duration of symptoms. Centres wishing to contribute to the Registry should contact Maria Loughenbury at maria.loughenbury@brit-thoracic.org.uk

The BTS ILD Registry is supported by Interstitial Lung Disease patient charities; both Action for Pulmonary Fibrosis and SarcoidosisUK have lay representation on the steering group. Lay summaries are available for the UK IPF Registry and the UK Sarcoidosis Registry, both available on the BTS website.

We believe that sharing data will ultimately improve patient care. We hope you find this report helpful and will continue to work alongside us.

***Professor Andrew Wilson***
***Chair, BTS Interstitial Lung Disease Registry Steering Group***

*Data is power and the BTS ILD Registry remains the established tool for understanding ILD in the UK, and over the past year its potential as a means to support research and service provision has also been more widely recognised.*

*Perhaps the most significant development this year is that of embedding the Registry in the NHSE ILD Quality Dashboard, providing assurance on the quality of patient care in England. BTS has been heavily involved in determining Dashboard metrics, which are easily collated through the UK IPF Registry. Although this is only a requirement for English specialist services, resources developed to facilitate this work are available to Registry participants across the UK, supporting centres within the devolved nations to benchmark themselves against these critical aspects of patient care.*

*Research is at the heart of improving patient care and, since the launch of the BTS Data Access Request Process last year, six successful applications have been made to access Registry data for research. This level of engagement is encouraging, especially considering these data have only been made available to researchers since the start of the COVID-19 pandemic (an incredibly challenging time for us all).*

*I look forward to the BTS ILD Registry evolving further, responding to the ever-changing UK respiratory landscape.*

**Professor Jonathan Bennett**
**Chair, BTS Board of Trustees (2019-2021)**

## BTS Lung Disease Registry Steering Group Membership 2021:

Professor Andrew Wilson, Chair
Dr Huzaifa Adamali, Consultant Respiratory Physician
Mrs Sarah Agnew, Nurse Representative
Mr Howard Almond, Patient Representative
Mr Leo Casimo, SarcoidosisUK
Dr Ahmed Fahim, Consultant Respiratory Physician
Dr Wendy Funston, Trainee Respiratory Physician
Dr Sarah Haney, Consultant Respiratory Physician
Professor Ling-Pei Ho, Consultant Respiratory Physician
Dr John Hutchinson, Consultant Respiratory Physician
Mr Steve Jones, Action for Pulmonary Fibrosis
Dr Philip Molyneaux, Consultant Respiratory Physician
Dr Katherine Spinks, Consultant Respiratory Physician

Miss Sally Welham, BTS Chief Executive
Mr Miguel Souto, BTS Head of Clinical Programmes
Miss Maria Loughenbury, BTS Lung Disease Registry Manager

# ACKNOWLEDGEMENTS

The BTS Lung Disease Registry Programme is funded by the British Thoracic Society. A grant (2012-2014) from the Healthcare Quality Improvement Partnership (HQIP) contributed to the initial development of the Interstitial Lung Disease Registry and this support is gratefully acknowledged.

The Society is grateful for financial assistance provided from Boehringer Ingelheim and InterMune for the enhancement of the data collection software (2014).

The BTS Interstitial Lung Disease Registry is supported by:

Action for Pulmonary Fibrosis – www.actionpf.org/

SarcoidosisUK – www.sarcoidosisuk.org

The British Lung Foundation – www.blf.org.uk

**Cover photograph**: Liz Robertson, a patient living with interstitial lung disease. With thanks to Action for Pulmonary Fibrosis.

# INTRODUCTION

The BTS ILD Registry was launched in February 2013 and includes two registries: the UK IPF Registry and the UK Sarcoidosis Registry.

The BTS ILD Registry was developed with the aim of improving standards of care for patients with IPF and sarcoidosis. This includes enabling and facilitating research to improve understanding of the epidemiology and progression of these diseases.

## Who can participate in the ILD Registry and how many are doing so now?

The Registry is open to all secondary and tertiary care institutions in England, Scotland, Wales and Northern Ireland. The data cut examined in this report was taken on 30th June 2021, at a time when 75 sites across 60 Trusts/Health Boards had obtained approval to participate. This current full list of 75 participating sites is given on page 33.

Overall the BTS ILD Registry includes over 4,100 patient records (3,385 IPF records and 774 sarcoidosis records).

## Data Entry

Three sets of data are collected for each individual patient:

- Patient demographic information (age, gender, comorbidities, etc.).
- Clinical features on diagnosis and at first clinic visit.
- Follow-up information from subsequent clinic visits (at 12 month intervals following entry onto the BTS ILD Registry).

Clinical information includes questions about disease behaviour, treatments given and referral to other key services, as well as capturing metrics in line with the published NICE IPF Quality Standard[1].

## Registry Ethics Approval, Information Governance and Data security

Ethical approval for the British Thoracic Society Interstitial Lung Disease Registry Programme (17/EE/0346) was granted by the NRES Committee East of England in October 2012. It was renewed in October 2017. Patient consent must be obtained before any patient information is entered into the BTS ILD Registry. Information for patients and copies of consent forms are available on the BTS website at:
https://www.brit-thoracic.org.uk/quality-improvement/lung-disease-registries/bts-ild-registry/.

Participating centres are required to confirm their Caldicott Guardian has approved their participation in the Lung Disease Registry before they can enter data.

All patient identifiable data (e.g. name, date of birth, postcode) are encrypted at the point of entry. Therefore, identifiable data can only be accessed by the hospital team directly responsible for caring for the patient. No patient identifiable data are available to BTS ILD Registry administrators. The British Thoracic Society Information Governance Policy and associated data security policy documents are available on the BTS website at:
https://www.brit-thoracic.org.uk/about-us/governance-documents-and-policies/

## Notes on data and percentages/denominators

Throughout this report figures are displayed as percentages and as exact figures (of the format *numerator/denominator*). Where figures are presented in the form *numerator/denominator* the denominator may vary for a number of reasons. Although the UK IPF Registry includes 3,385 patient demographic records, not all questions are fully completed by centres for every patient. For example, family history was recorded for only 2,545 records and thus the denominator in this case would be 2,545.

When reading this report please be aware that:

- Denominators in this report always exclude cases where no response was entered;
- Unless otherwise stated, denominators in this report exclude cases where the saved response was ‘not known’ or ‘not recorded’; and
- Percentage figures are rounded to the nearest whole number throughout this report. This means rounding errors may lead to some total percentages adding up to 99% or 101%.

# PART 1 – The Impact of the BTS ILD Registry

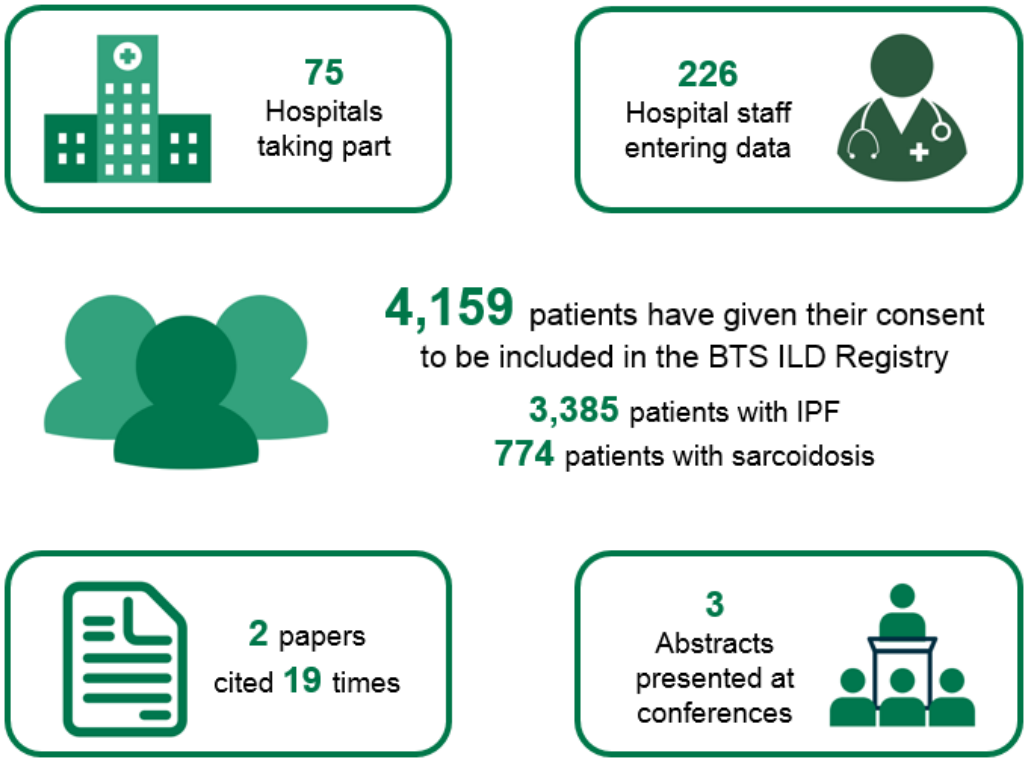
This report is based on data representing eight and a half years of hard work and dedication from those involved in the ILD Registry Steering Group and from the hundreds of Registry collaborators who have collected and entered data voluntarily.

The BTS ILD Registry impacts on the care of patients with IPF and sarcoidosis by:

- **Increasing understanding** of the clinical and disease burden by providing an annual overview of Registry data and publishing findings.

- **Facilitating the delivery of care** in a number of ways:
  - Enabling services to benchmark their own local delivery against national delivery;
  - Supporting English specialist centres to complete NHSE ILD Quality Dashboard returns;
  - Helping hospital management to monitor their service against key standards;
  - Providing tools to assist with administering patients locally; and
  - Ultimately, helping to driving improvements in patient care across the UK.

- **Facilitating research** in a number of ways:
  - Identifying willing study participants (at the time consent is taken patients are asked if they would like to be contacted should any suitable trial become available);
  - Partnering with research projects to assist with data collection;
  - Capturing enrolment in research, allowing this to be monitored over time; and
  - Providing a mechanism for independent research using Registry data, through the BTS Data Access Request Process (https://www.brit-thoracic.org.uk/quality-improvement/bts-clinical-data-policy-and-data-access/).



# PART 2 – The UK Idiopathic Pulmonary Fibrosis Registry

Idiopathic pulmonary fibrosis (IPF) is a chronic, progressive interstitial disease with a dismal prognosis. The symptom burden experienced by patients with IPF is high, with the majority of patients presenting with breathlessness, cough and fatigue. Some individuals may also experience anxiety and depression. There remain significant delays from symptom onset to the diagnosis, and survival remains poor; since the Registry launched in 2013 the reported all-cause mortality rate is 35%,

It has been a challenging year for medical community, and the respiratory workforce in particular has been deployed to manage the increasing numbers of inpatient admissions with COVID-19 since the start of 2021. This is reflected in a reduction of entries to the UK IPF Registry in the first six months of 2021, with just 64 entries with the patient first presenting at the clinic in the first half of 2021 compared to 245 entries with the patient first presenting in the first half of 2019.

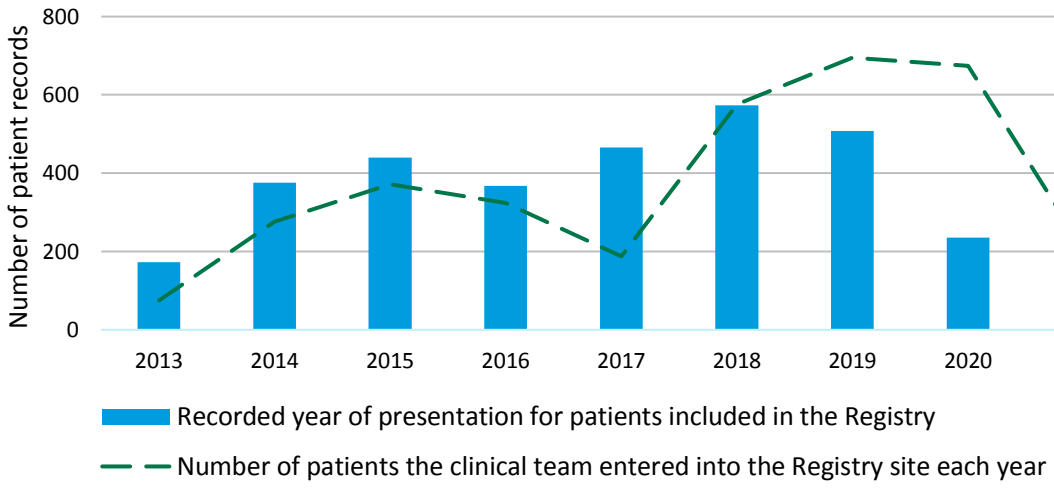



**Figure 1**: **UK IPF Registry participation over time**
The number of patients with data entered onto the Registry over time, and the year of presentation for those patients. Data may be entered both prospectively and retrospectively, therefore the year of data entry is not always the year of presentation. This chart clearly shows the impact of the COVID-19 pandemic on data entry in 2020/21 – reduced data entry, with an increased delay in submitting patient records. Data for 2021 represent the first six months of the year only.

Historically very few patients have been enrolled into clinical trials. It is exciting that UK IPF Registry is being used to collect data for use in the randomised Treating Idiopathic Pulmonary Fibrosis with the Addition of Lansoprazole (TIPAL) clinical trial. This collaboration between the UK IPF Registry and the Norwich Clinical Trials Unit (which is running the study) means that lung function and other relevant data are captured directly from the Registry, without double data entry into case report forms. All other trail data are captured remotely with minimal involvement from sites. This study is open for recruitment from all sites working with the Registry which are, or are associated with, UK ILD Specialist Centres. For more information contact tipal@uea.ac.uk

This report also presents data against each of the Quality Statements in the Quality Standard for Idiopathic Pulmonary Fibrosis[1] as published by the National Institute for Clinical Excellence (NICE). This is particularly timely in view of the imminent mandatory reporting of data from English specialist centres

to the NHSE Interstitial Lung Disease (ILD) Quality Dashboard, which is primarily centred around those Quality Statements.

BTS has been working alongside NHSE since for a number of years to update the NHSE ILD Quality Dashboard. The new Dashboard items are designed to be easy for centres to capture and to provide a true representation of local delivery and standards of care. Full details of the updated NHSE ILD Quality Dashboard items are included on the BTS website, along with details of how all centres can use the resources provided to benchmark their delivery (the Dashboard is mandated for English specialist centres, but district general hospitals and hospitals from the devolved nations are very welcome to use these resources). For more information please contact registry@brit-thoracic.org.uk

The report shows that a high proportion of individuals are diagnosed only following consensus at a Multidisciplinary Team (MDT) Meeting and large proportion of individuals have assessment for oxygen, pulmonary rehabilitation referral and palliative care needs. However, only three quarters of patients have the opportunity to interact with a specialist ILD nurse, highlighting a gap in service provision that should be addressed.

The data show that ant-fibrotic prescriptions remain stable over time, with approximately 44% of patients being prescribed an antifibrotic drug. Moreover, the proportion of other pharmacological therapies – including proton pump inhibitors (PPIs) – remain fairly similar to the previous years.

There are several other highlights from this report including:

- There remains a long delay from symptom onset to diagnosis, with 40% of patients waiting for more than 2 years.
- There continues to be a decline in the number of lung biopsies to confirm diagnosis (from 15% in 2013 to 2.5% in 2021). However, these data are likely to be skewed in 2020-2021 due to the COVID-19 pandemic and cessation of elective diagnostic surgical procedures.
- The percentage of patients discussed in ILD MDT remain stable at above 90%.
- The stable, high burden of comorbidities with 84% with at least one comorbidity.
- The majority of patients remain in GAP I and II categories (99%), as before.
- Increased palliative care needs assessments (from 74% in 2013 to 91% in 2019, with data from 2020 (82%) and 2021 (95%) highly variable due to the COVID-19 pandemic).

This year, a paper prepared by Dr Helen Parfrey and colleagues, analysing the UK IPF Registry dataset from 2013-19, was published on behalf of the British Thoracic Society[2] and in 2020 Dr Phil Molyneaux and colleagues investigated the consequences of using different algorithms for determining predicted lung function values, and the cost implications of changing to the Global Lung Index [3].

Researchers from external organisations can also investigate outcomes from the Registry using the BTS Data Access Request Process (for more details please visit https://www.brit-thoracic.org.uk/quality-improvement/btsclinical-data-policy-and-data-access).

## Inclusion criteria

Participating centres are asked to enter data on patients who meet the following inclusion criteria:

- Patients with definite or strongly suspected idiopathic pulmonary fibrosis.
- Patients with a new diagnosis of IPF made at a clinic visit from 1st January 2013 onwards.
- Patients with a historical diagnosis of IPF seen for the first time in the clinic at the participating centre from 1st January 2013.

Patients must provide written consent before their data may be entered into the UK IPF Registry.

Patients with non-idiopathic disease (e.g. those with a history of significant asbestos exposure, strong possibility of sub-clinical or evolving connective tissue disease, or clear history of exposure to drugs or antigens known to cause interstitial lung disease) are not eligible for inclusion in the UK IPF Registry.

Data may be entered both prospectively and retrospectively. When entering retrospective data, as long as the patient's first clinic visit was on or after 1st January 2013 all of their historical information – from their first visit and each of their follow-up visits – may be entered into the Registry.

## Available data to 30th June 2021

At the end of June 2021, 75 centres had approval to participate in the UK IPF Registry, with 55 centres having contributed clinical data. The full list of 75 centres which have approval to participate in the Registry is given on page 33.

**3,385** patient demographic records – an increase of 588 patients compared to the end of June 2020

**2,684** complete clinical/diagnosis records from the first clinic visit

**2,624** Follow-up records representing 1,098 unique patients. These follow-up records represent 41% (1,098/2,684) of all complete records, with patients where follow-up data have been entered having a mean of two follow-up records each.

## 2.1 THE IPF PATIENT COHORT

Patient demographic information is collected at the first clinic visit.

### Gender

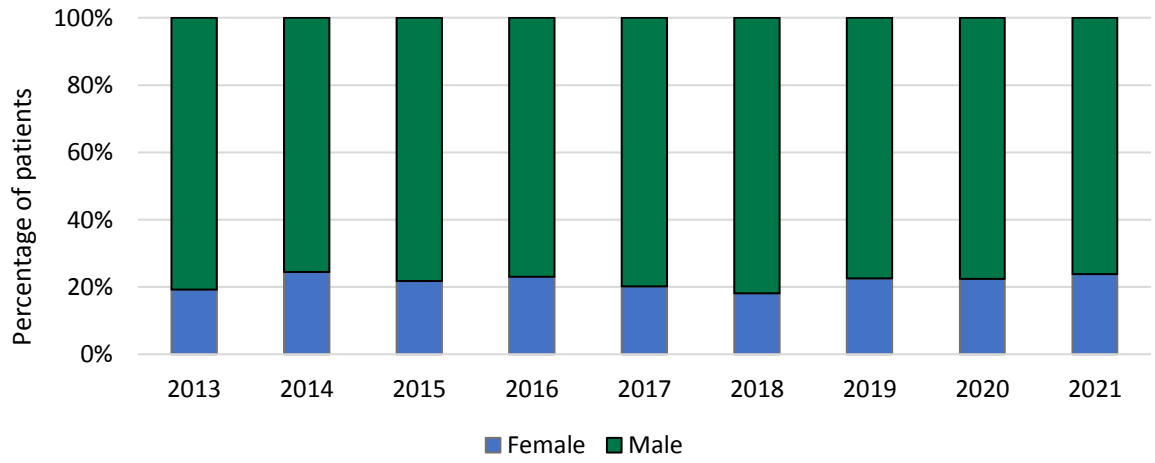



**Figure 2**: **Referrals by gender over time**
The proportion of male and female patients has remained consistent over time, with a mean of 78.5% (2,572/3,275) males and 21.5% (703/3,275) females over the lifetime of the Registry. These data are in keeping with the known epidemiology of this disease.

### Age

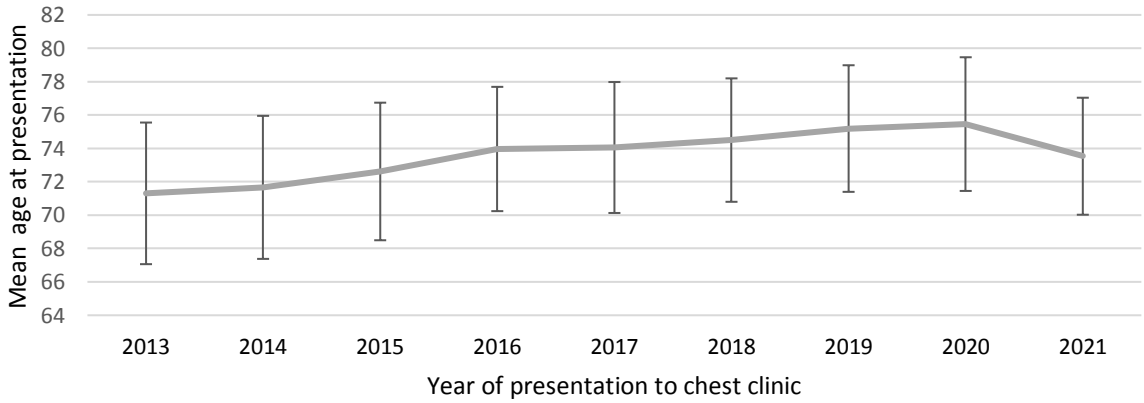



**Figure 3**: **Mean age at presentation over time**
The mean age of patients at presentation (shown here with error bars representing standard deviation) gradually increased over time from 71.3 years in 2013 to 75.5 years in 2020. However, in 2021 the mean age was lower at 73.5 years. This may represent the referral pattern during the COVID-19 pandemic.

Over the lifetime of the Registry the mean age was 73.7 (± *8.0*), with 72% (2,339/3,244) aged 70 or over.

## Comorbidities

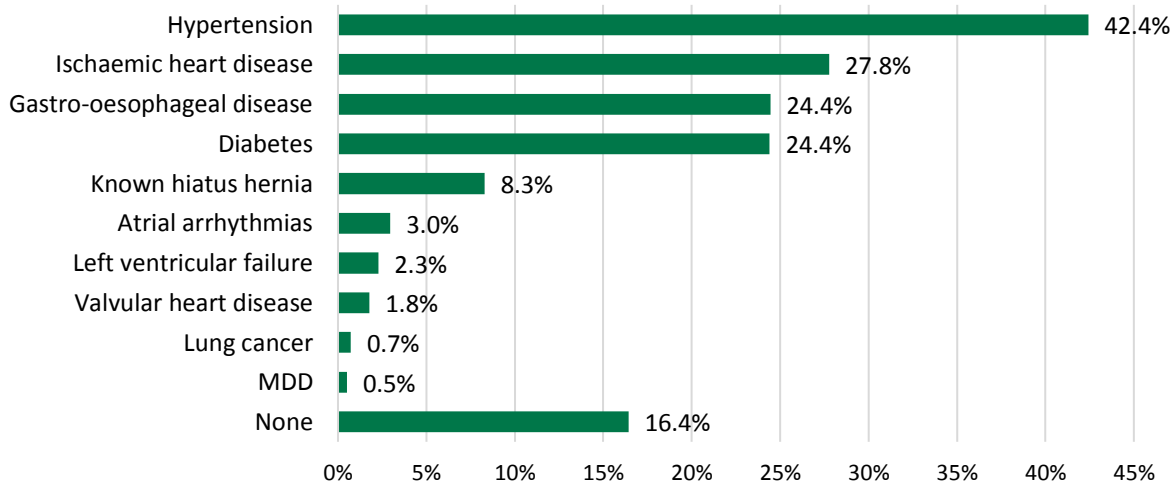



**Figure 4: Comorbidities at presentation**
Patients with IPF have a high burden of comorbidities at presentation, with 84% having at least one comorbidity. The nature and proportion of comorbidities remain stable over the lifetime of the Registry.

## Smoking

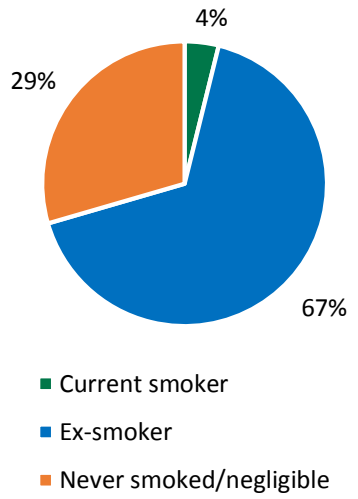



**Figure 5: Smoking status at presentation**
The proportion of patients who were smokers at presentation has remained consistent through the lifetime of the Registry. Overall, 4% (101/2,605) of patients were current smokers, 67% (1,736/2,605) ex-smokers and 29% (768/2,605) had never smoked or smoked a negligible amount (<5 pack years).

## Relatives with IPF

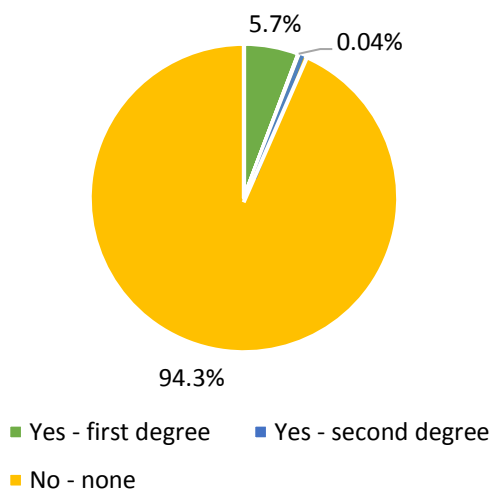



**Figure 6: Known relatives with IPF**
Where clinicians had recorded family history, approximately 5.7% (145/2,545) of patients had a first degree relative known to have IPF and 94.3% (2,399/2,545) had no known relatives with IPF. These figures remained relatively consistent throughout the lifetime of the Registry.

## 2.2 DIAGNOSING IPF

The data presented in this section were collected once, at the first clinic visit.

### Duration of chest symptoms prior to presentation

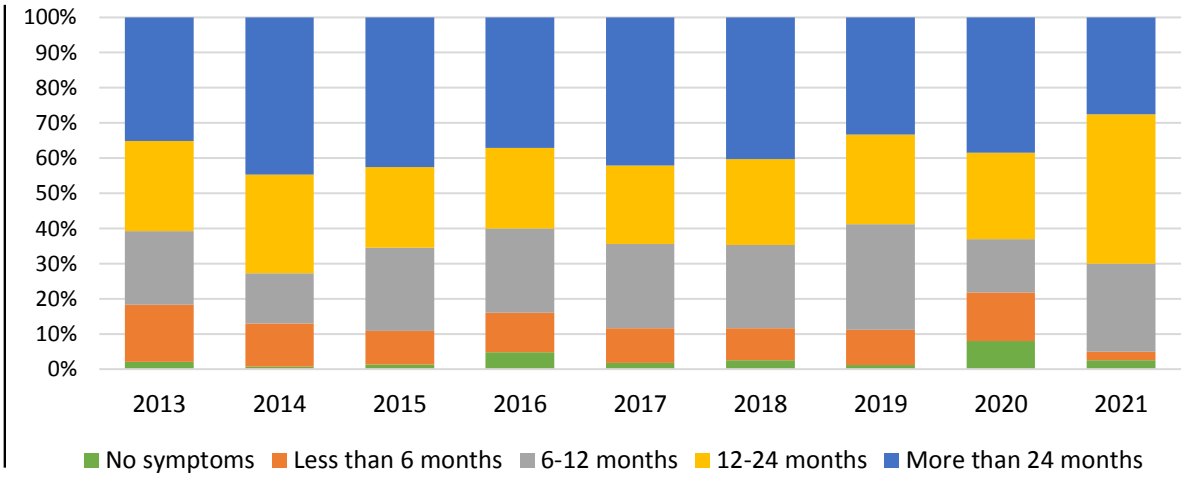



**Figure 7**: **Duration of IPF symptoms prior to presentation over time**

Unfortunately, patients often have symptoms for a considerable period prior to diagnosis. Since 2013, 63.5% (1,619/2,551) of patients reported having chest symptoms for more than 12 months before their first hospital clinic visit. The time from symptom onset to diagnosis seems to have been stable over time until the COVID-19 pandemic in 2020. Data from 2021 include only 6 months (to June).

### Route of referral

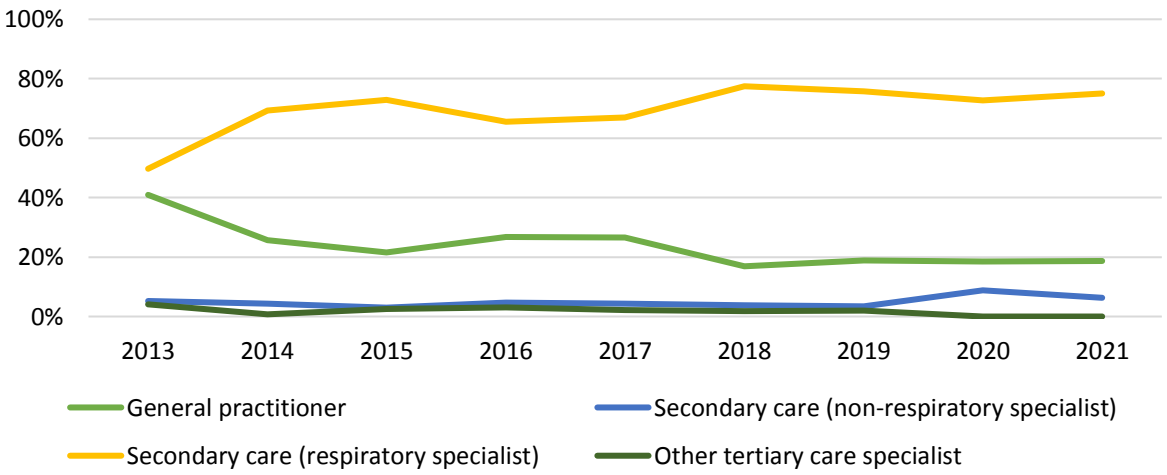



**Figure 8**: **Route of referral to clinic over time**

Throughout the lifetime of the UK IPF Registry the primary route of referral has been directly from a respiratory specialist in secondary care. Since 2013 there appears to have been a significant increase in referral from respiratory specialists in secondary care (from 49% in 2013 to 75% in 2021, although only 6 months of data are available for 2021) and a comparable reduction in referral from general practice (from 41% in 2013 to 19% in 2021). This may be due to the introduction of antifibrotic therapy, for which referral to tertiary care is required.

## Patient Waiting Times

Mean waiting times for patients with IPF from referral to first clinic visit were 13.4 weeks (13.6 for English specialist centres and 11.9 for other centres).

Mean waiting times from referral to multidisciplinary team (MDT) meetings were 11.5 weeks (11.3 and 12.5 for English specialist centres and other centres respectively).

## High-Resolution Computed Tomography (HRCT) and Surgical Lung Biopsy

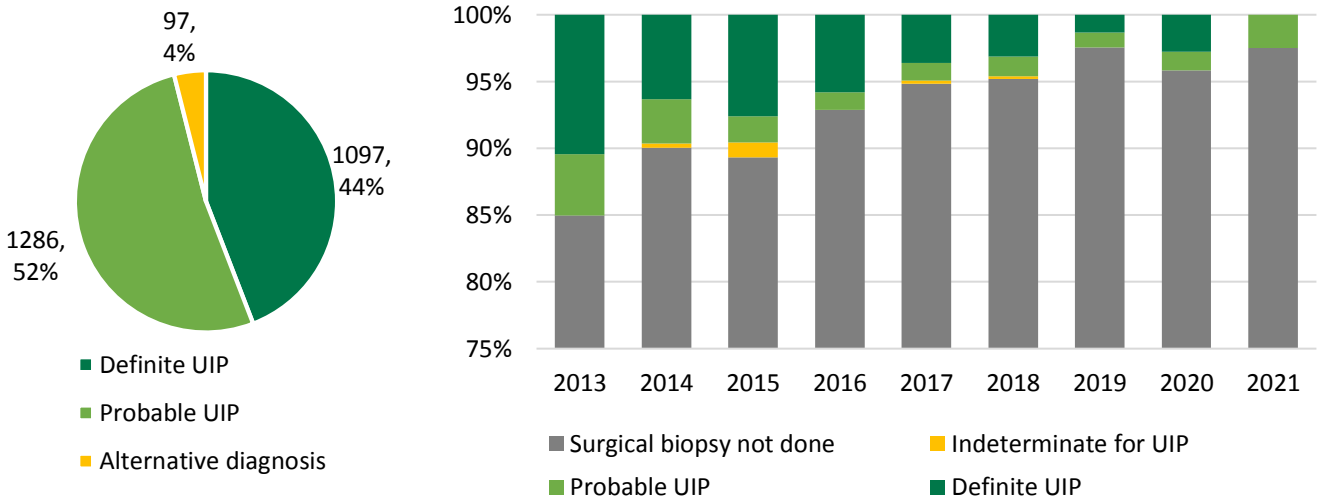



**Figure 9 (left)**: **HRCT pattern at presentation, and**
**Figure 10 (right)**: **Surgical biopsy data at presentation over time**
The majority of patients are diagnosed using high-resolution computed tomography (HRCT) scanning. Roughly equal proportions of these scans reported patterns in keeping with definite and probable usual interstitial pneumonia (UIP), which is an important factor in the diagnosis of IPF. Relatively few patients require a biopsy to confirm the diagnosis (7%, 184/2,589). The percentage of cases where a biopsy was conducted decreased from 15% in 2013 to 2.5% in 2021, especially since the diagnostic guidelines were updated in 2018. Data for 2021 represent only the first six months of the year (January to June).

## MDT

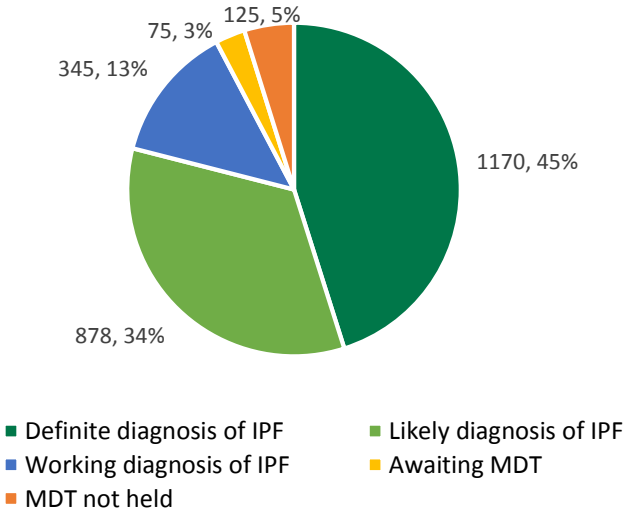



**Figure 11**: **Outcome of MDT**
The majority of cases are discussed at MDT (with 92% having been discussed at the first clinic visit and a further 3% due to be discussed at an upcoming MDT).

There appears to have been a steady fall in the number of cases not being discussed at MDT, reducing from 15.0% (21/141) of cases in 2013 to 4% of cases in 2020 (6/149) – see page 36.

## 2.3 CLINICAL DATA AT PRESENTATION

The data presented in this section are collected once for each patient, at the first clinic visit.

### GAP Staging

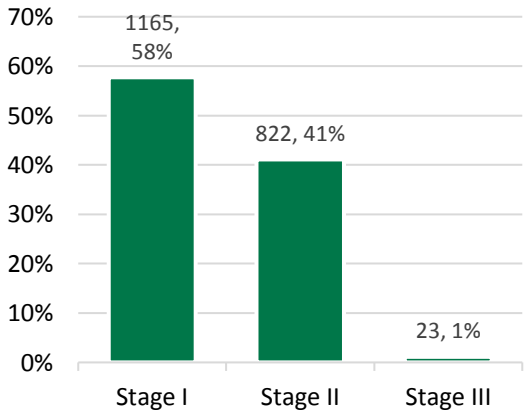



**Figure 12**: **GAP stage at presentation**
At first presentation the overwhelming majority of patients with IPF are categorised as having GAP Stage I or Stage II disease (99% of patients overall – 1,987/2,010). GAP staging is a marker of IPF disease severity, calculated using gender, age and lung function details[4].

### Pulmonary hypertension

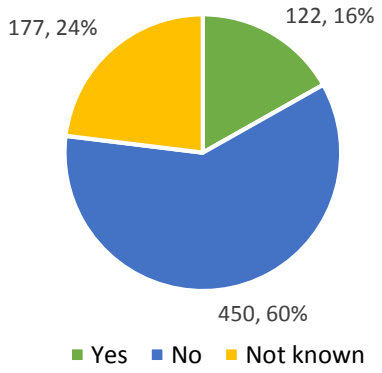
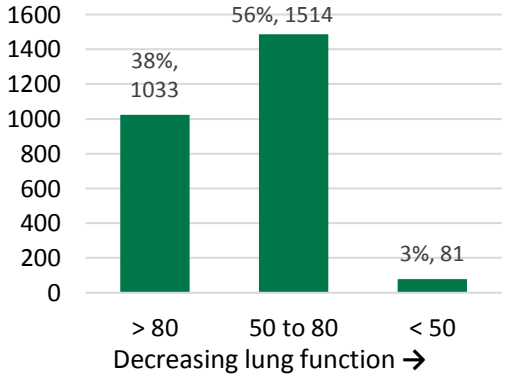



**Figure 13**: **History of pulmonary hypertension**
Overall 16% (122/749) of patients at presentation had previously been confirmed to have pulmonary hypertension or right heart strain, secondary to their lung disease, confirmed on echocardiogram or right heart catheterisation. This question was added in December 2019, therefore the denominator of 749 is comparatively low.

### Lung Function (at presentation)




| FVC Grouping | No. of patients | Mean DLCO % predicted | Standard Deviation |
|---|---|---|---|
| < 50% | 81 | 37.8% | *15.0* |
| 50 to 80% | 1,514 | 47.0% | *20.5* |
| > 80% | 1,033 | 55.4% | *17.9* |

**Figure 14 (left)**: **Forced vital capacity (FVC) at presentation, and**
**Table 1 (right)**: **DLCO and FVC at presentation over time**
At entry 38% of patients have an FVC over 80% predicted and are therefore above current England's National Institute for Health and Care Excellence (NICE) criteria for antifibrotic treatment. However, UK IPF Registry data show these patients already exhibit a substantial decrease in their diffusing capacity for carbon monoxide (mean DLCO= 55%).

In England, Wales and Northern Ireland patients with FVC values <50% predicted are less likely to be referred on to a specialist centre, as treatment cannot be accessed and/or they may be too unwell to travel. This likely explains why only 3% of patients on the UK IPF Registry have lower FVC values.

## Drug treatment at presentation

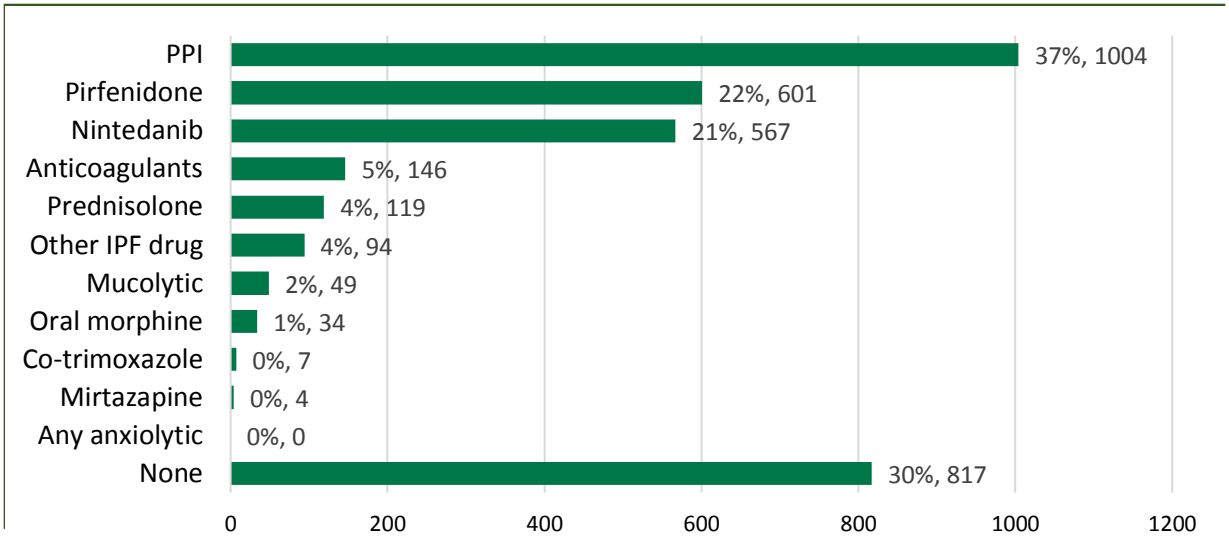



**Figure 15**: **Drug treatment at presentation (including treatment prescribed at first clinic visit)**
These data need to be interpreted carefully as they reflect drug use over the lifetime of the UK IPF Registry. Pirfenidone has been available through the NHS since 2013, whereas nintedanib has only been available since 2016. Registry questions were also amended in December 2019 to include mirtazapine and mucolytic as new answer options, meaning these will appear artificially low.

## Reasons for not Prescribing Antifibrotic Treatment

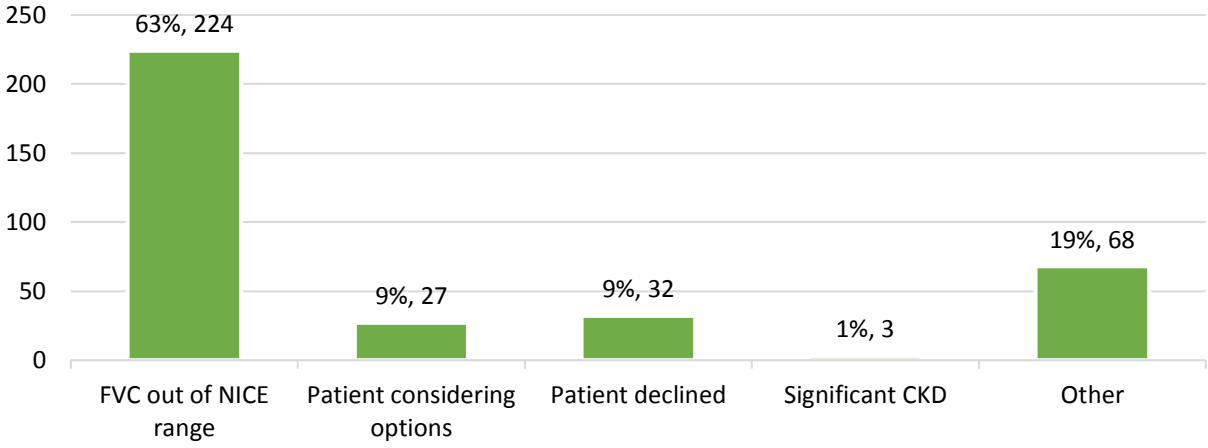



**Figure 16**: **Reason for not starting antifibrotic treatment at first clinic visit**
Since 2013, 55% of patients were not prescribed antifibrotic treatments at presentation. Suitability for drug treatment is not based purely on the FVC treatment criteria defined by NICE. Other factors, such as renal or liver function abnormalities, can sometimes preclude use of drugs.

From December 2019 clinicians were asked why patients were not receiving antifibrotic therapy. For almost two thirds of patients (63%) this was because the FVC was outside the range approved by NICE. For only 9% of patients this was because they spent time after their clinic visit considering their treatment options. Some participating centres are also not able to prescribe antifibrotic treatment, and future reports will make a greater distinction between data from prescribing and non-prescribing sites.

## Oxygen Therapy

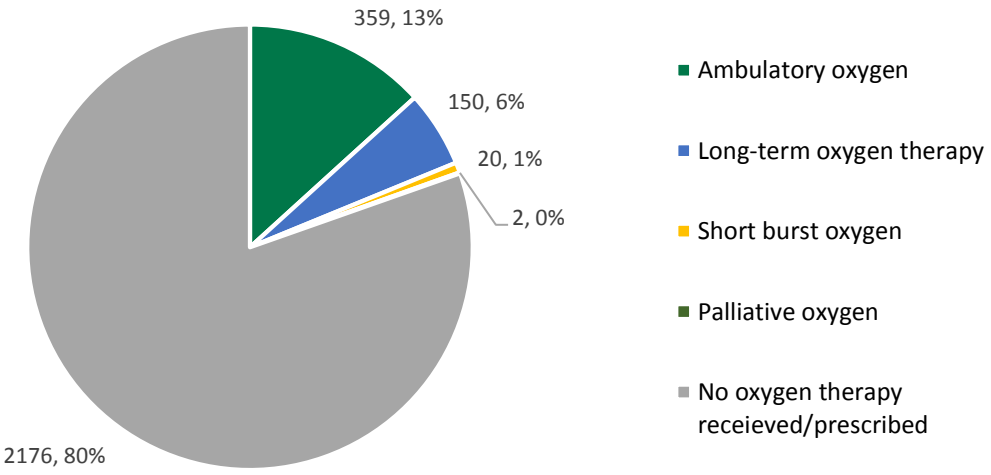



**Figure 17**: **Oxygen therapy at presentation**
At presentation 94% (578/613) of patients had their oxygen needs assessed, with the majority (72%, 414/578) not requiring oxygen therapy at that time – see page 38. One in six (17%, 445/2,621) patients were receiving or newly prescribed at least one form of oxygen therapy at their first clinic visit.

## Lung Transplantation

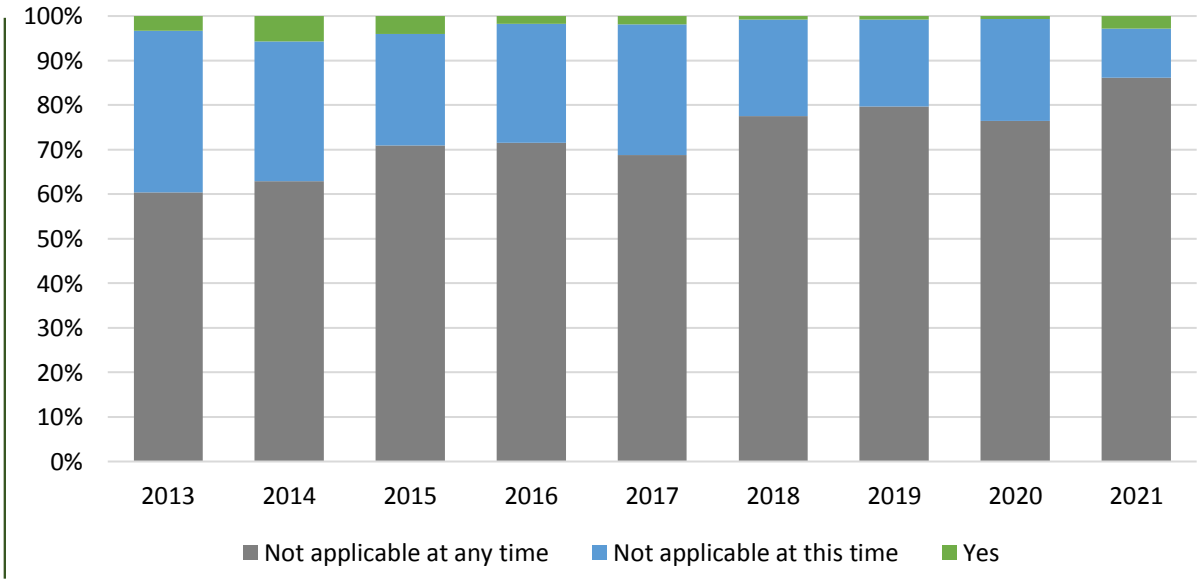



**Figure 18**: **Referral for lung transplantation at presentation over time**
Over the lifetime of the UK IPF Registry the proportion of patients deemed ineligible for lung transplantation 'at any time' at their first clinic visit appears to have increased gradually, rising from 60% in 2013 to 86% in 2021 (data for 2021 represent only the first six months of the year). There are a number of possible reasons for this, including the gradual increase in age at presentation over time (see Figure 3, page 13).

## Inclusion in Clinical Trials

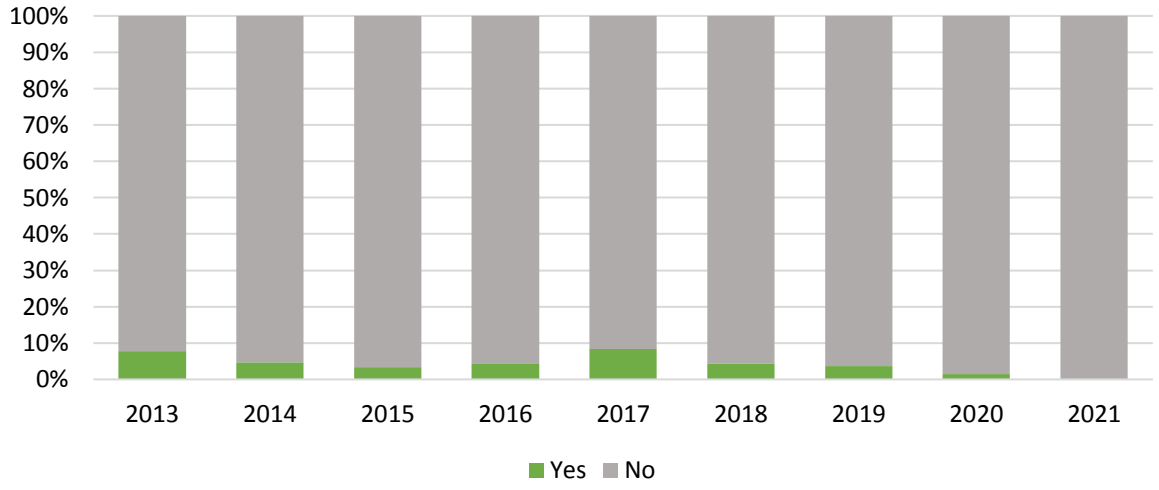



**Figure 19**: **Inclusion in clinical trials at presentation over time**
The proportion of patients recruited to clinical trials at the time of presentation has remained consistently low throughout the lifetime of the UK IPF Registry. Data from 2021 represent only the first six months of the year.

## Patient Mortality

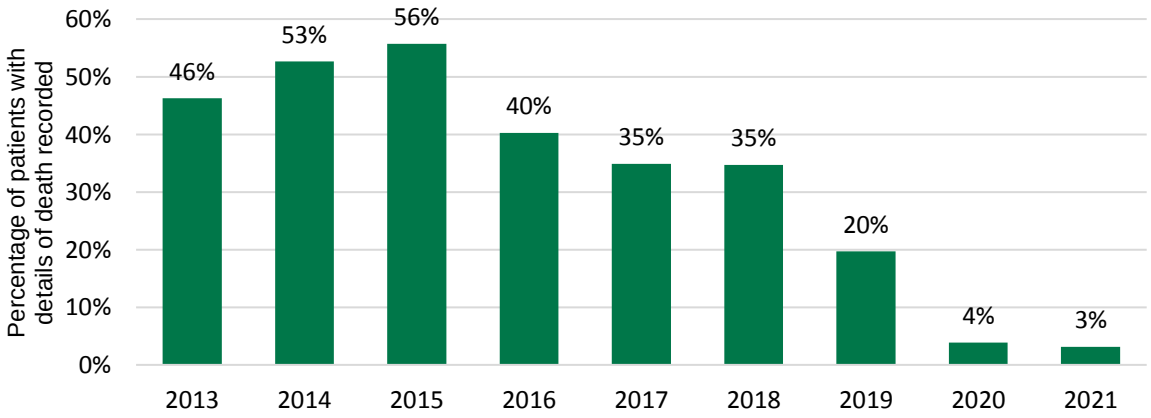
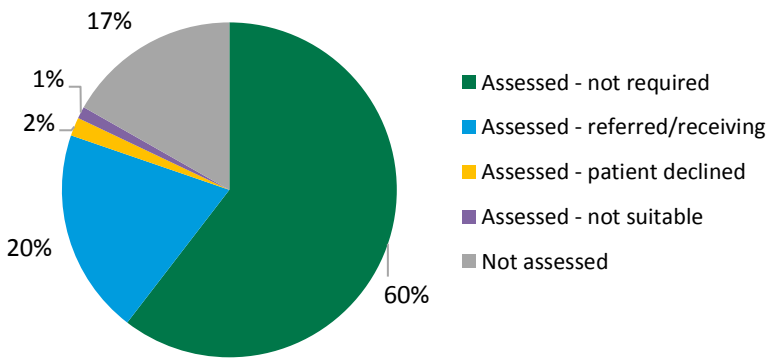



**Figure 20**: **Patient mortality by year of presentation**
The overall IPF mortality rate for the lifetime of the Registry is 35%. This chart breaks that figure down, showing the mortality rates to date of patients who first presented to the treating centre each year. These data should be interpreted with caution for a number of reasons, including:

- The patients included in the Registry are overwhelmingly (approximately 90%) entered from specialist, prescribing centres. This means the population is skewed toward patients who have less mild disease and are within the antifibrotic treatment for English specialist centres.
- The Registry dataset holds data on patient mortality, but participating sites can only enter this information if they are aware of it themselves. If centres have not specifically been informed of a patient passing (e.g. directly informed, or identifying this when preparing to arrange a follow-up clinic visit) there would be a delay before mortality information would be completed.

## 2.4 NICE QUALITY STANDARD FOR IPF

In this section, data from the UK IPF Registry are presented in relation to the five Quality Statements in the NICE Quality Standard for IPF[1].

| IPF Quality Statements | |
|---|---|
| **Quality Statement 1:**<br><br>**People are diagnosed with IPF only with the consensus of a multidisciplinary team (MDT) with expertise in interstitial lung disease.** | This standard appears to be being met in the majority of cases, with UK IPF Registry data showing 92% of cases have already been discussed at MDT by the time of the first clinic visit and a further 3% due to be discussed at an upcoming MDT. See Figure 11, page 16, and see Item 1, page 36. |
| **Quality Statement 2:**<br><br>**People with IPF have an interstitial lung disease specialist nurse available to them.** | In December 2019 a question was added to the UK IPF Registry, asking if the patient had been offered the opportunity to see or provided contact details for an ILD specialist nurse at presentation.<br><br>At presentation 77% (482/627) of patients were offered the opportunity to interact with an ILD specialist nurse (see Item 2, page 36). Ideally this figure would be 100%. |
| **Quality Statement 3:**<br><br>**Patients with IPF have an assessment for home and ambulatory oxygen therapy at each follow-up appointment and before they leave hospital following an exacerbation of the disease.** |  <br>**Figure 21**: **Oxygen assessment at annual review**<br>Clinicians are asked if they have addressed and managed the oxygen needs of the patient at follow-up clinic visits. Overall 83% (793/953) of follow-up visits included an oxygen needs assessment. Of those assessed, the majority did not require oxygen therapy at the time (73%, 576/793). This question was added to the UK IPF Registry dataset in December 2019, therefore the number of responses is comparatively low. See Item 6, page 38. |

**Quality Statement 4:**

**Pulmonary rehabilitation (PR) programmes provide services that are designed specifically for IPF.**

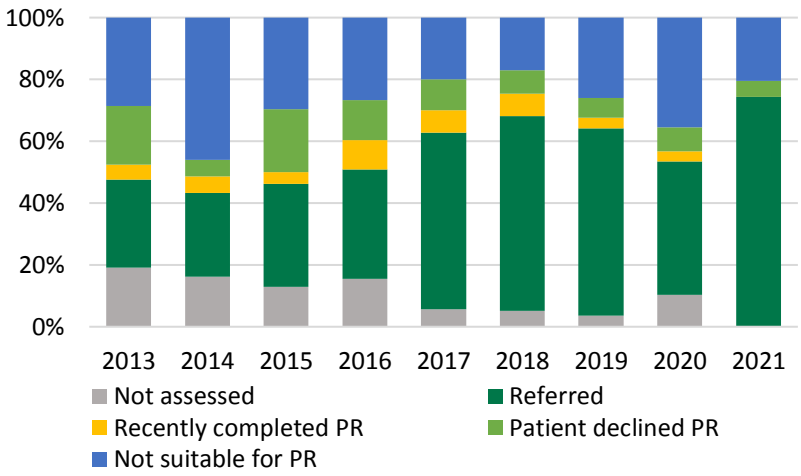



**Figure 22**: **PR needs assessment at presentation over time**

The UK IPF Registry does not hold data on whether PR services are designed specifically for patients with IPF; however, it does hold information regarding PR needs assessment. At presentation, 93% (1,382/1,483) of patients had their PR needs assessed and 7% (101/1,483) did not. Of those assessed the majority (60%, 818/1,382) were referred for PR.

There appears to have been an increase in the proportion of patients referred for PR over time, from 18% in 2013 to 54% in 2019 (with data varying widely in 2020/1 due to the COVID-19 pandemic). These data should be interpreted with caution, as data regarding PR have only been collected in this form since January 2017. Consequently, there are many fewer records containing PR data prior to 2017 (being limited to retrospective data only).

There were a number of reasons given for patients not being referred for PR, including the patient declining or having recently (within the last twelve months) completed a course of PR. Of all patients whose PR needs were assessed, 25% (347/1,382) were thought not to be suitable for referral, either due to poor mobility or already having a good fitness level. See Item 3, page 37.

**Quality Statement 5:**

**People with IPF and their families and carers have access to services that meet their palliative care needs.**

At presentation 83% (1,254/1,503) patients were reported to have had their palliative care needs assessed. See Item 4, page 37. By palliative care we mean care intended to optimise quality of life and reduce suffering (for example, through symptom management).

When completing this question, clinicians are advised that the patient's needs have still been assessed even if that assessment identifies no current need for palliative support.

There appears to have been a gradual increase in palliative needs assessment at presentation over the lifetime of the Registry, from 74% in 2013 to 91% in 2019. However, as above, the COVID-19 pandemic lef to greater variability in 2020 (82%) and 2021 (95%).

# PART 3 – The UK Sarcoidosis Registry

Sarcoidosis is a multisystem condition characterised by granulomatous inflammation which can occur in any organ. Although the lungs are most frequently affected (over 90% of patients), other sites such as the skin, eye, joints, nervous system, liver, spleen, muscles, nose and sinuses are involved to varying extents. The inflammation commonly resolves without sequalae but, in a proportion of patients, it results in progressive irreversible scarring. The heterogeneous nature of the condition means that it can present in a variety of manners and with varied symptom severity, and that patients can present to clinicians from a variety of specialist areas. However, the most common symptoms are cough and fatigue.

Much the same as for the UK IPF Registry, the referral patterns, disease management and Registry data entry have all altered since the start of the COVID-19 pandemic. Therefore, comparisons with recent data and previous years should be treated with caution.

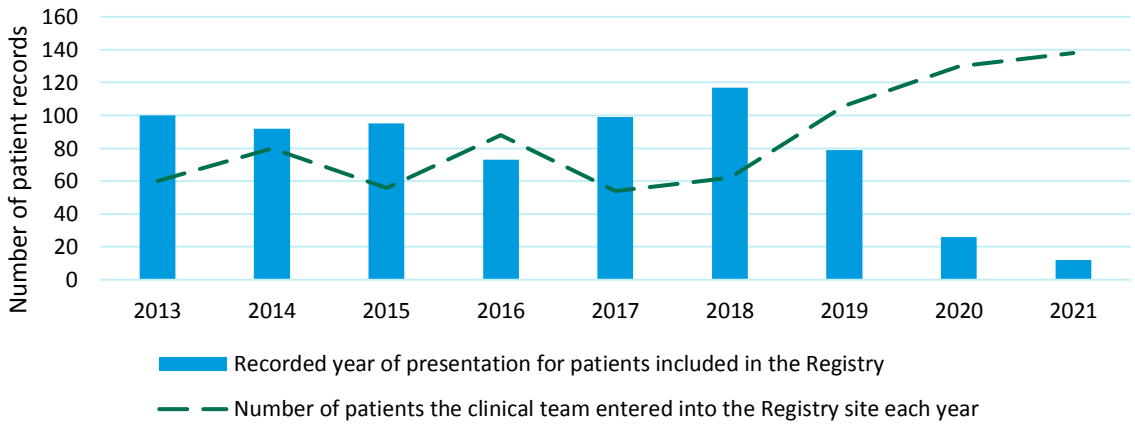



**Figure 23**: **UK Sarcoidosis Registry participation over time**
The number of patients with data entered onto the Registry over time, and the year of presentation for those patients. Data may be entered both prospectively and retrospectively, therefore the year of data entry is not always the year of presentation. This chart clearly shows that data entry to the Registry is increasing, but that there are comparatively few records where patient data was entered in 2021/2 – this is likely due to an increase in retrospective data entry, possibly as centres were able to see fewer patients in clinic as a result of the COVID-19 pandemic. There were just 38 entries with the patient first presenting at the clinic in the 18 months from January 2020 to June 2021, compared to 48 entries with the patient first presenting in the first half of 2019.

The UK Sarcoidosis Registry holds data captured by respiratory physicians, and it is hosted by the British Thoracic Society. As such, the majority of patients have respiratory involvement, with only 7% of patients having normal HRCT scans. Although this reflects our understanding of this condition, patients with mild disease or those not having respiratory involvement may be underrepresented in this database. Likewise, only 7% of patients recorded on the Registry were black, although we know the prevalence of sarcoidosis is up to 4 times more common in black populations [5] and these individuals may be underrepresented in the database.

Data collected from 41 centres over the lifetime of the Registry (January 2013 to the end of June 2021) indicate that sarcoidosis predominantly affects women and presents most commonly between the ages

of 41 to 60 years (52% of patients). The majority of people were never smokers with only 8% currently smoking. Most patients were free from significant comorbidities but had a high symptom burden with 84% of people having at least one symptom.

Other highlights from these data include:

- Over the duration of the Registry Endobronchial ultrasound (EBUS) examination has gradually replaced mediastinoscopy and surgical lung biopsy. It was a recorded method of tissue sampling in 71% of cases in 2019 (data in 2020 and 2021 are highly variable due to the COVID-1 pandemic).
- 10% of patients are receiving immunosuppressive therapies (other than prednisolone).
- Only 3% of patients are recruited into clinical trials
- Few patients (31%) are provided information about other services, such as patient support groups, helplines or mental health services.

## Inclusion criteria

Participating centres are requested to enter data on patients who meet the following inclusion criteria:

- Patients with a new diagnosis of sarcoidosis made at a clinic visit from 1st January 2013 onwards.
- Patients with a historical diagnosis of sarcoidosis seen for the first time in the clinic at the participating centre from 1st January 2013.

Data may be entered both prospectively and retrospectively. When entering retrospective data, as long as the patient's first clinic visit was on or after 1st January 2013 all of their historical information – from their first visit and each of their follow-up visits – may be entered into the Registry.

## Available data to 30th June 2021

At the end of June 2021, 75 centres had approval to participate in the UK Sarcoidosis Registry, with 41 centres having contributed clinical data. The full list of 75 centres which have approval to participate in the Registry is given on page 33.

**774** patient demographic records

**518** complete clinical/diagnosis records from the first clinic visit

**250** follow-up records representing 139 unique patients.

## 3.1 THE SARCOIDOSIS PATIENT COHORT

Patient demographic information is collected at the first clinic visit.

### Gender

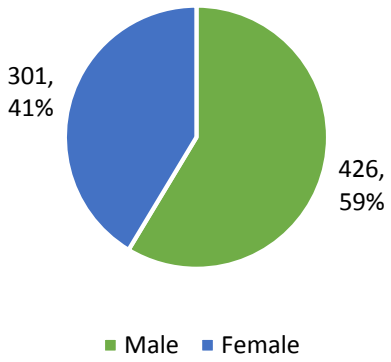



**Figure 24**: **Gender of patients**
The proportion of male (59%, 426/727) and female (41%, 301/727) patients has remained consistent over the lifetime of the UK Sarcoidosis Registry.

### Age

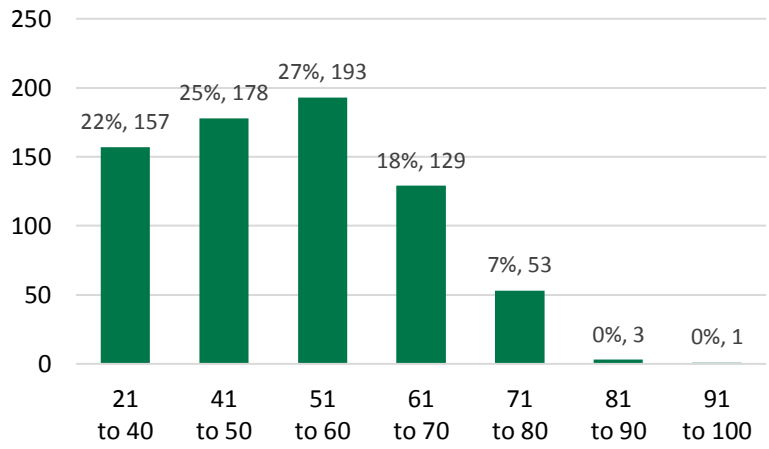



**Figure 25**: **Age at presentation**
Just over a quarter of patients (26%, 186/714) presented over the age of 60, with the mean age at presentation being 51.5 years, with a standard deviation of ± 13.2.

### Ethnicity

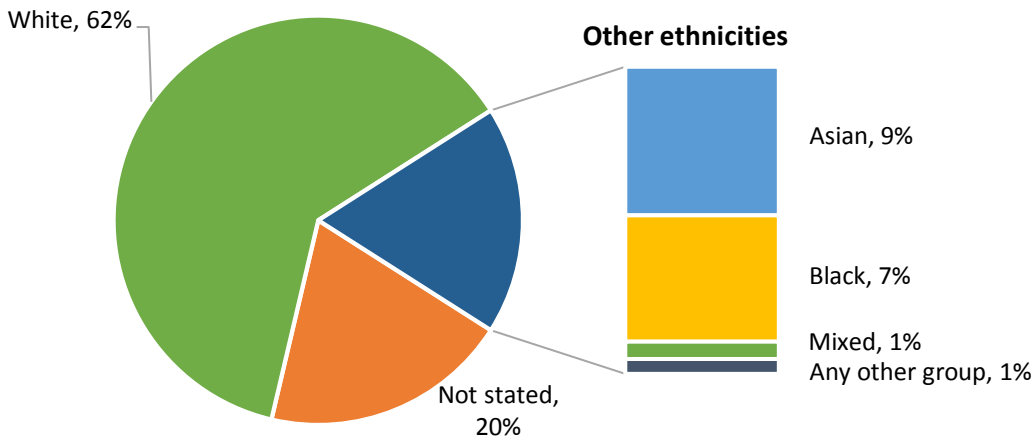



**Figure 26**: **Patients by ethnic group**
The majority of patients in the Registry were white (62%, 435/696). Although sarcoidosis in known to be more prevalent in black populations, only 7% (51/696) of patients were black. This figure likely reflects the populations from which Registry data were obtained, as UK census data from 2011 indicate 86% of the population of England and Wales reported their ethnicity as White, whereas only 3% described themselves as Black/African/Caribbean/Black British[6].

## Smoking status

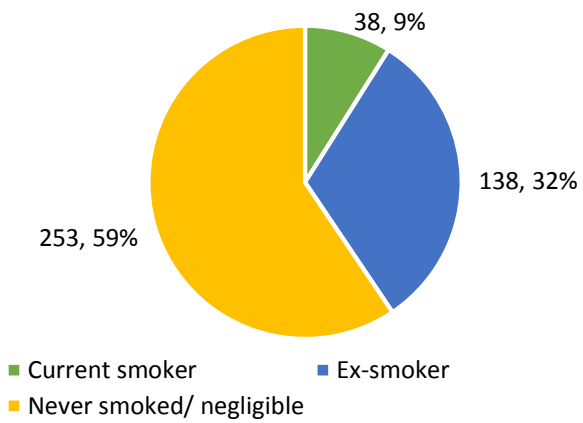



**Figure 27**: **Smoking status at presentation**
Overall 41% (176/429) of patients were either smokers or ex-smokers at presentation (patients were counted as ex-smokers if they quit more than three months before their first clinic visit).

59% (253/429) of patients had either never smoked or only smoked a negligible amount (defined as less than five pack years).

## Comorbidities

Almost two thirds of patients (66%, 234/357) had no reported comorbidity at the time of their current presentation

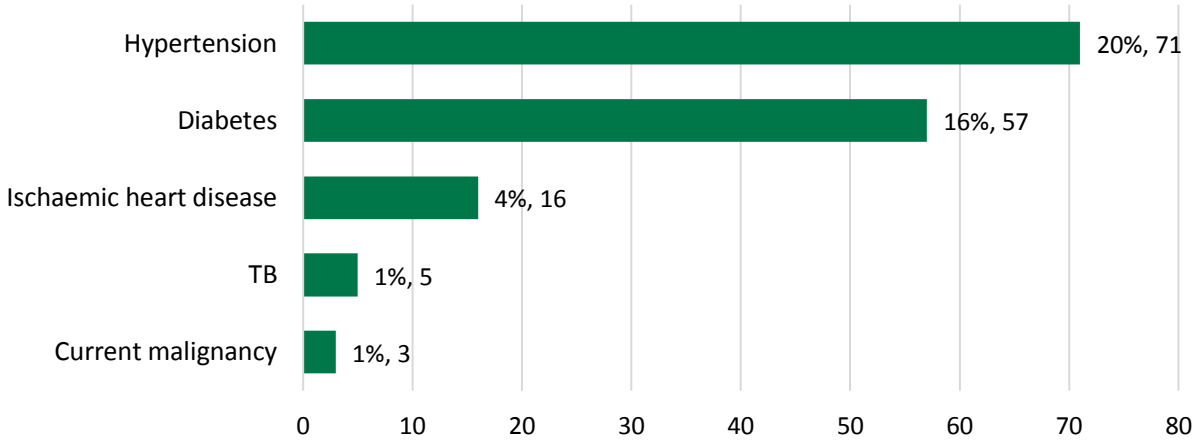


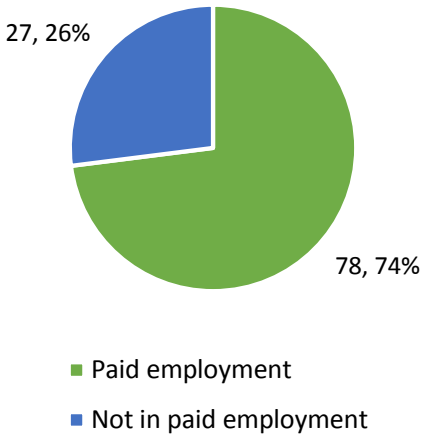

**Figure 28**: **Patient comorbidities at presentation**
The most commonly reported comorbidities were systemic hypertension (one in five patients – 20%, 71/357) and diabetes (15%, 57/357). These conditions are highly prevalent in the general population. Where at least one comorbidity was recorded patients had a mean of 1.2 reported comorbidities each.

These figures differ from previous reports because data are collected against fewer comorbidities since the UK Sarcoidosis Registry dataset was updated in December 2019.

## Relatives with sarcoidosis

A minority of patients in the UK Sarcoidosis Registry are known to have relatives who have been diagnosed with sarcoidosis. Overall, 4% (16/438) of patients reported having at least one first degree relative previously diagnosed with sarcoidosis.

## Employment and burden of disease




In December 2019 the UK Sarcoidosis Registry dataset up was updated to include questions on employment status. As more data are collected over time it is anticipated that these questions will allow for a greater understanding of the burden of disease nationally.

**Figure 29**: **Employment status at presentation**
Where employment status at presentation was known, almost three quarters (74%, 78/105) of patients were in paid employment. Of those who were not in paid employment the majority (81%) were retired.

## 3.2 DIAGNOSING SARCOIDOSIS

The data presented in this section were collected once, at the first clinic visit.

### Referral to clinic

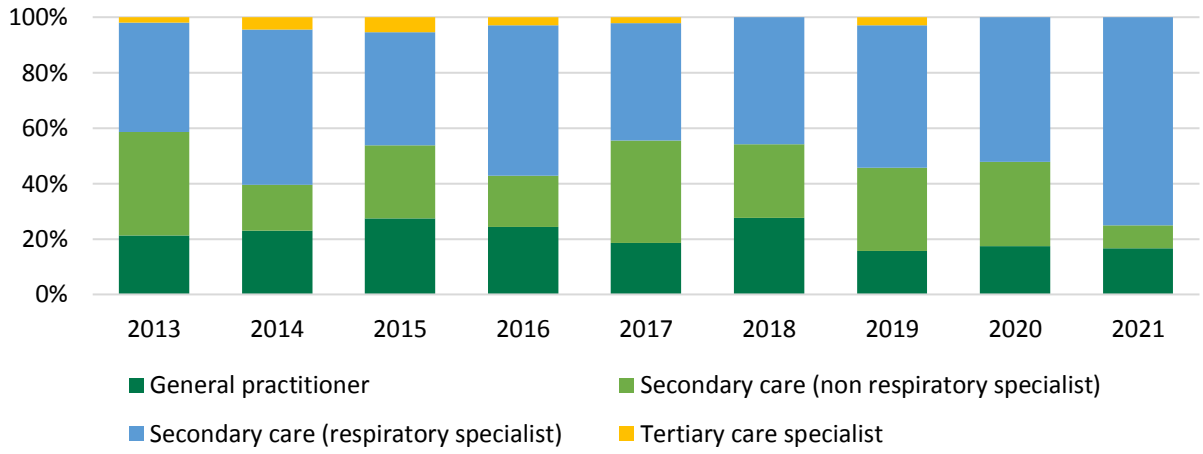



**Figure 30**: **Route of referral to clinic over time**

Overall, 47% (319/678) of patients were referred from respiratory physicians in secondary care. Referrals from general practice have remained low at 23% overall (155/678), and no higher than 28% in any given year. This may reflect lack of awareness in primary care and/or complexities in diagnosing sarcoidosis – as is evidenced by 37% (177/473) of cases known to have been diagnosed incidentally. Data from 2021 should be interpreted with caution, as they represent only the first six months of the year (to the end of June).

### Symptoms at first clinic visit

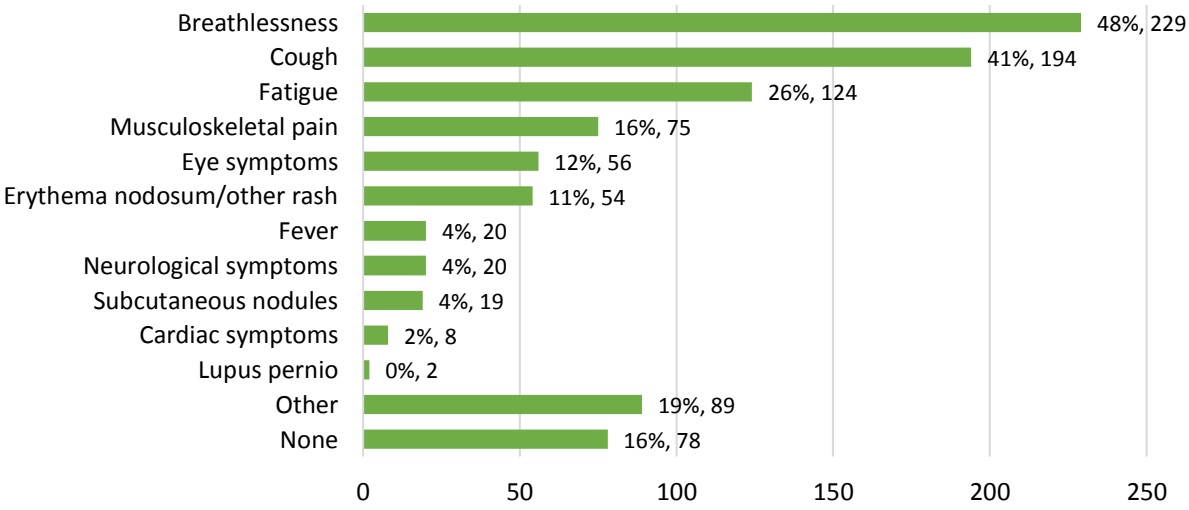



**Figure 31**: **Symptoms reported at first clinic visit**

The most common symptoms were breathlessness (48%), cough (41%) and fatigue (26%). Musculoskeletal pain (16%), eye symptoms (12%) and skin rashes (11%) were also frequently reported. Almost one in six (16%) patients had no symptoms recorded at first clinic visit.

## Diagnostic biopsies

Over the lifetime of the UK Sarcoidosis Registry 95% (349/369) of patients had at least one biopsy conducted during their diagnostic investigations. However, it should be noted that patients who receive a biopsy may be more likely to have their details entered onto the Registry.

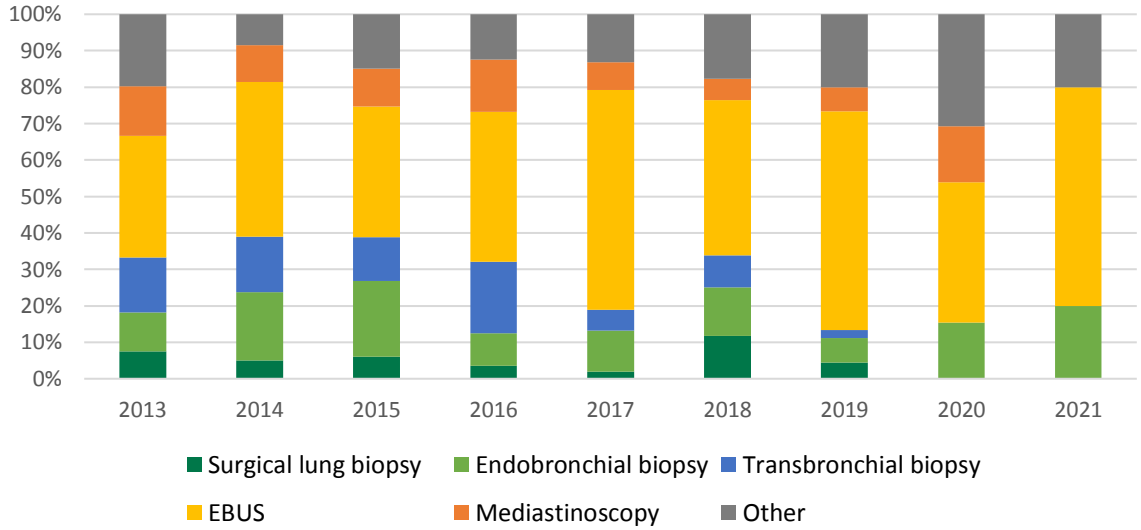



**Figure 32**: **Biopsy techniques used at presentation over time**
A number of different biopsy techniques were employed. Endobronchial ultrasound (EBUS) was consistently the most popular, involved in 55% (191/349) of cases where at least one biopsy was conducted. Data from 2021 should be interpreted with caution, as they represent only the first six months of the year.

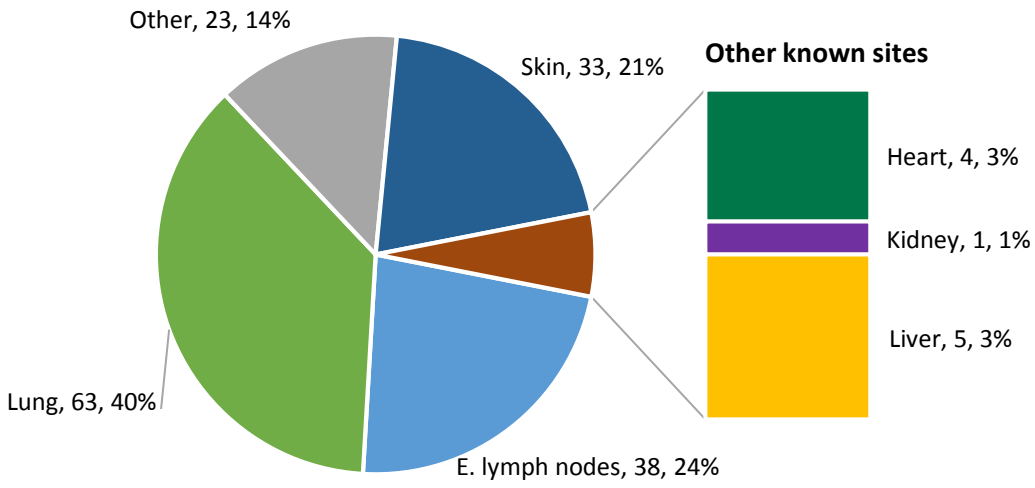



**Figure 33**: **Site(s) from which histology obtained at presentation**
The most common biopsy sites were lung (40%, 63/159), extra-thoracic lymph nodes (24%, 38/159) and skin (21%, 33/159). The variety of biopsy sites investigated is indicative of the multisystem involvement commonly observed in sarcoidosis.

## 3.3 CLINICAL DATA AT PRESENTATION

The data presented in this section are collected once for each patient, at the first clinic visit.

### Pulmonary hypertension

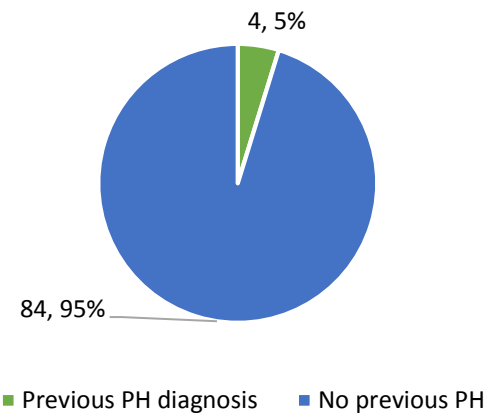



**Figure 34**: **History of pulmonary hypertension (PH)**

At presentation 5% (4/88) of patients had previously been confirmed to have pulmonary hypertension or right heart strain, secondary to their lung disease, confirmed on echo or right heart catheter from any hospital. This question was added to the UK Sarcoidosis Registry dataset in December 2019.

### MRC dyspnoea scale

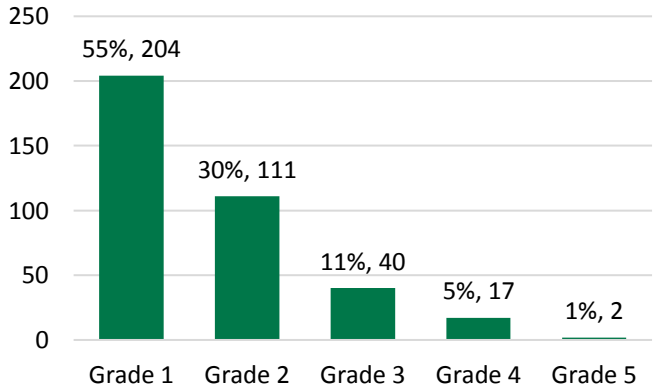



**Figure 35**: **Breathlessness at presentation**

The majority of patients (84%, 315/374) experienced mild or negligible shortness of breath at the time of presentation, having either Grade I (not troubled by breathlessness except on strenuous exercise) or Grade II (short of breath when hurrying or walking up a slight hill) breathlessness at presentation according to the Medical Research Council (MRC) dyspnoea scale.

### Chest radiograph

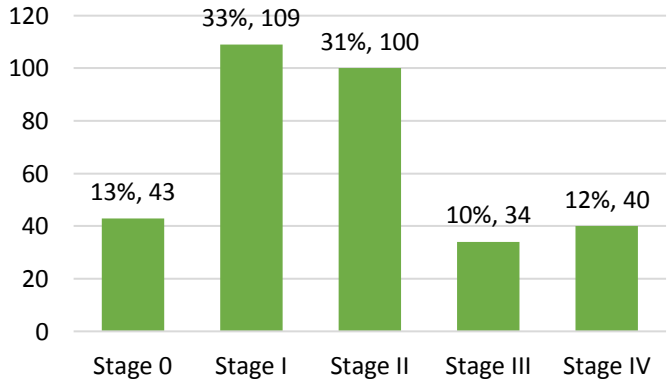



**Figure 36**: **Chest radiograph at presentation**

At presentation the majority of patients were observed to have either Stage I with lymphadenopathy only (33%, 109/326) or Stage II lymphadenopathy and parenchymal involvement (31%, 100/326) disease according to the Siltzbach sarcoidosis classification system[7].

## Blood tests

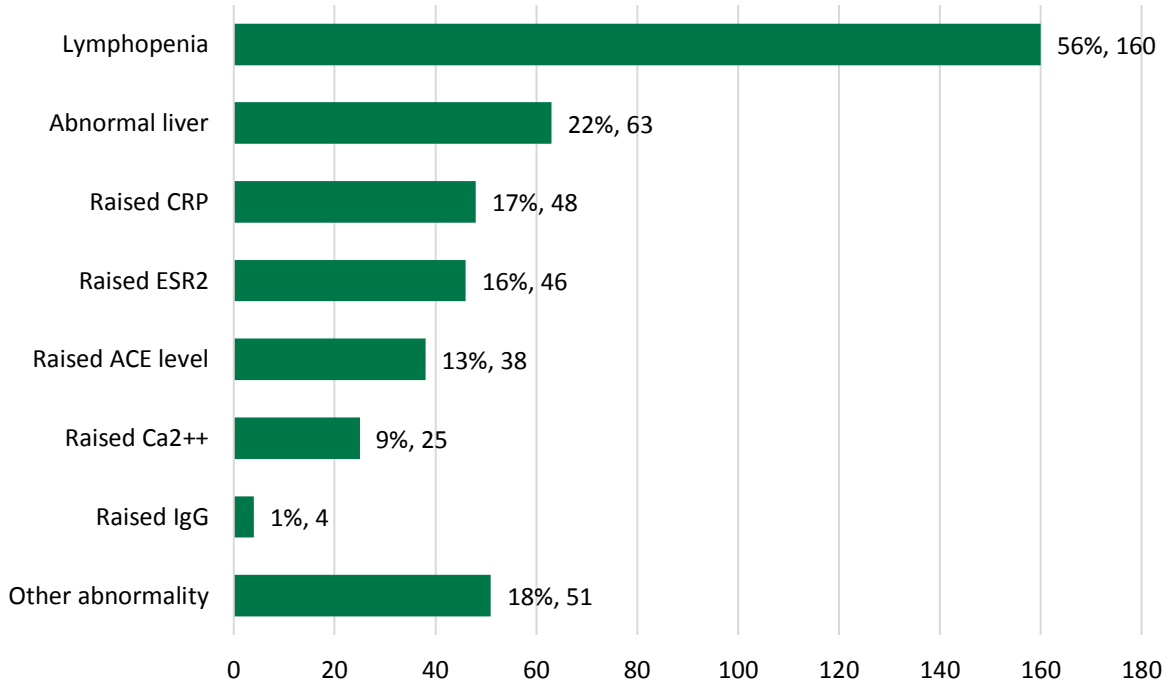



**Figure 37**: **Blood test abnormalities recorded at presentation**
The most common abnormality recorded in blood tests at presentation was lymphopenia, identified in 56% (160/285) of patients. Raised angiotensin converting enzyme (ACE) levels and calcium levels (as defined at local centres) were reported in 13% and 9% of cases respectively.

## HRCT pattern

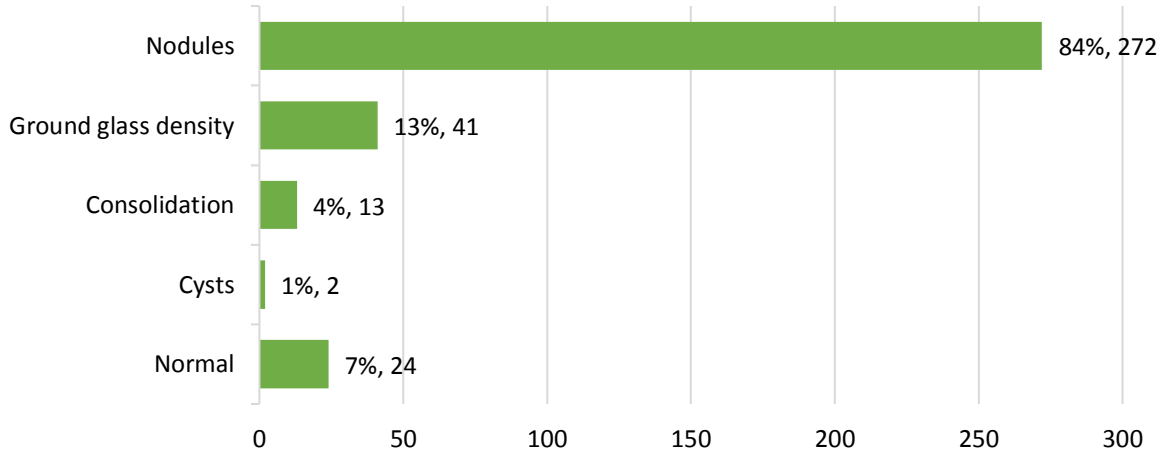



**Figure 38**: **Parenchymal abnormalities identified on HRCT at presentation**
The most common parenchymal abnormality identified on HRCT imaging at presentation was nodules, found in 84% (272/322) of cases. HRCT imaging was found to be normal in 7% (24/322) of cases.

## Current drug treatment

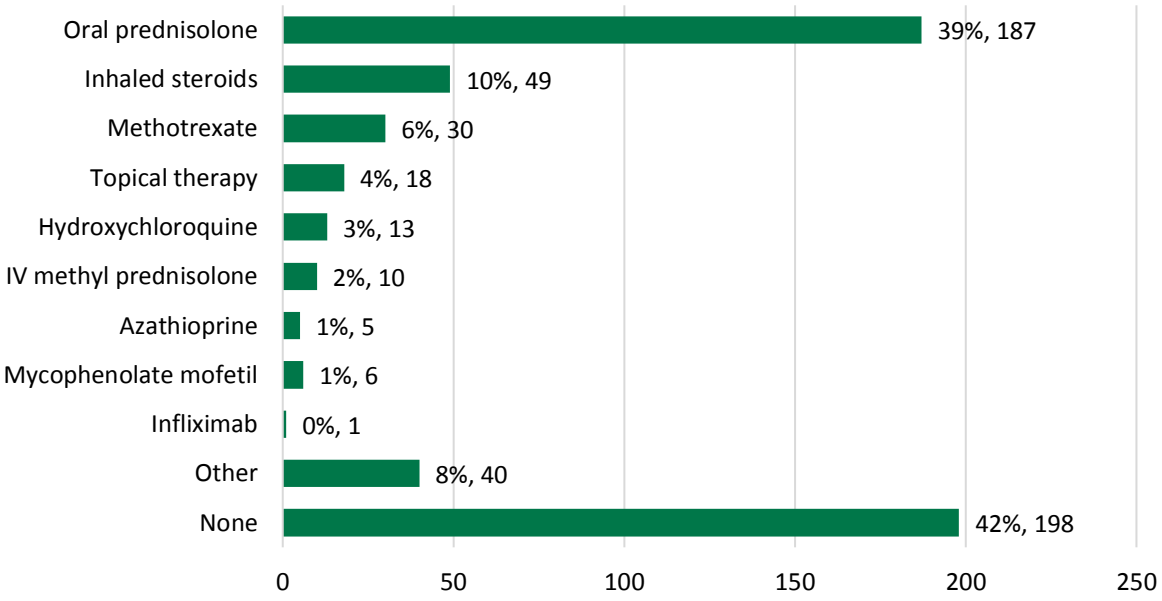



**Figure 39**: **Drug treatment at presentation**
The majority of patients were either not started on treatment (42%, 198/475) or managed with systemic corticosteroids (47%, 223/475). A number of alternative agents were used, with none used in more than 10% of cases. This broadly reflects previous BTS guidance on the management of sarcoidosis[8].

## Referral to other services

In December 2019 a question was added to the UK Sarcoidosis Registry dataset to determine what proportion of patients were signposted to other services. By other services we mean services providing either other clinical support (e.g. mental health services) or non-clinical support (e.g. support groups).

Early data indicate that only 31% (19/62) of patients were referred or signposted to other services at the time of presentation. Of those who were, 32% (6/19) were given details for patient support groups and 26% (5/19) for helplines, and 11% (2/19) were referred to or informed of mental health support.

## Inclusion in clinical trials

At presentation only 2.7% (12/445) of patients were recruited to a clinical trial. There is a need for more clinical research in sarcoidosis.

# PARTICIPATING SITES

The following organisations are currently participating in the BTS Interstitial Lung Disease Registry – our thanks to all involved:

## England

Addenbrooke's Hospital, Cambridge University Hospitals NHS Foundation Trust
Aintree University Hospital, Liverpool University Hospitals NHS Foundation Trust
Birmingham Heartlands Hospital, University Hospitals Birmingham NHS Foundation Trust
Blackpool Victoria Hospital, Blackpool Teaching Hospitals NHS Foundation Trust
Burnley General Teaching Hospital, East Lancashire Hospitals NHS Trust
Castle Hill Hospital, Hull University Teaching Hospitals NHS Trust
Central Middlesex Hospital, London North West University Healthcare NHS Trust
Cheltenham General Hospital, Gloucestershire Hospitals NHS Foundation Trust
Chorley and South Ribble Hospital, Lancashire Teaching Hospitals NHS Foundation Trust
Churchill Hospital, Oxford University Hospitals NHS Foundation Trust
City Hospital, Sandwell and West Birmingham NHS Trust
Countess of Chester Hospital, Cheshire and Wirral Partnership NHS Foundation Trust
Croydon University Hospital, Croydon Health Services NHS Trust
Darlington Memorial Hospital, County Durham and Darlington NHS Foundation Trust
Ealing Hospital, London North West University Healthcare NHS Trust
George Eliot Hospital, George Eliot Hospital NHS Trust
Glenfield Hospital, University Hospitals of Leicester NHS Trust
Gloucestershire Royal Hospital, Gloucestershire Hospitals NHS Foundation Trust
Good Hope Hospital, University Hospitals Birmingham NHS Foundation Trust
Guy's Hospital, Guy's and St Thomas' NHS Foundation Trust
Hammersmith Hospital, Imperial College Healthcare NHS Trust
Harrogate District Hospital, Harrogate and District NHS Foundation Trust
Hexham General Hospital, Northumbria Healthcare NHS Foundation Trust
Hinchingbrooke Hospital, North West Anglia NHS Foundation Trust
King's College Hospital, King's College Hospital NHS Foundation Trust
King's Mill Hospital, Sherwood Forest Hospitals NHS Foundation Trust
Liverpool Heart and Chest Hospital, Liverpool Heart and Chest Hospital NHS Foundation Trust
Musgrove Park Hospital, Somerset NHS Foundation Trust
New Cross Hospital, The Royal Wolverhampton NHS Trust
Norfolk and Norwich University Hospital, Norfolk & Norwich University Hospitals NHS Foundation Trust
North Devon District Hospital, Northern Devon Healthcare NHS Trust
Northern General Hospital, Sheffield Teaching Hospitals NHS Foundation Trust
North Middlesex University Hospital, North Middlesex University Hospital NHS Trust
North Tyneside General Hospital, Northumbria Healthcare NHS Foundation Trust
Northwick Park Hospital, London North West University Healthcare NHS Trust
Nottingham City Hospital, Nottingham University Hospitals NHS Trust
Royal Papworth Hospital, Royal Papworth Hospital NHS Foundation Trust
Peterborough City Hospital, North West Anglia NHS Foundation Trust
Queen Alexandra Hospital, Portsmouth University Hospitals NHS Trust
Queen Elizabeth Hospital, University Hospitals Birmingham NHS Foundation Trust
Queen Elizabeth Hospital, Gateshead Health NHS Foundation Trust
Royal Blackburn Teaching Hospital, East Lancashire Hospitals NHS Trust
Royal Brompton Hospital, Royal Brompton and Harefield NHS Foundation Trust
Royal Derby Hospital, University Hospitals of Derby & Burton NHS Foundation Trust
Royal Devon and Exeter Hospital, Royal Devon & Exeter Foundation NHS Trust
Royal Free Hospital, Royal Free London NHS Foundation Trust
Royal Lancaster Infirmary, University Hospitals of Morecambe Bay NHS Foundation Trust
Royal Preston Hospital, Lancashire Teaching Hospitals NHS Foundation Trust

Royal Victoria Infirmary, The Newcastle upon Tyne Hospitals NHS Foundation Trust
Russells Hall Hospital, The Dudley Group NHS Foundation Trust
Solihull Hospital, University Hospitals Birmingham NHS Foundation Trust
Southampton General Hospital, University Hospital Southampton NHS Foundation Trust
Southmead Hospital, North Bristol NHS Trust
St James' University Hospital, The Leeds Teaching Hospitals NHS Trust
St Mary's Hospital, Imperial College Healthcare NHS Trust
University College Hospital, University College London Hospitals NHS Foundation Trust
University Hospital, University Hospitals Coventry & Warwickshire NHS Trust
University Hospital of North Midlands, University Hospitals of North Midlands NHS Trust
University Hospital of North Tees, North Tees & Hartlepool NHS Foundation Trust
Wansbeck Hospital, Northumbria Healthcare NHS Foundation Trust
Worcester Royal Hospital, Worcestershire Acute Hospitals NHS Trust
Wythenshawe Hospital, Manchester University NHS Foundation Trust

## Scotland

Aberdeen Royal Infirmary, NHS Grampian
Forth Valley Royal Hospital, NHS Forth Valley
Glasgow Royal Infirmary, NHS Greater Glasgow and Clyde
Lorn & Islands District General Hospital, NHS Highland
Ninewells Hospital, NHS Tayside
Perth Royal Infirmary, NHS Tayside
Royal Alexandra Hospital, NHS Greater Glasgow and Clyde
Vale of Leven District General Hospital, NHS Greater Glasgow and Clyde

## Wales

Glan Clwyd Hospital, Betsi Cadwaladr University Health Board
University Hospital Llandough, Cardiff and Vale University Health Board
Wrexham Maelor Hospital, Betsi Cadwaladr University Health Board

## Northern Ireland

Antrim Area Hospital, Northern Health and Social Care Trust
The Ulster Hospital, South Eastern Health and Social Care Trust

If you would like to know more about the BTS Interstitial Lung Disease Registry please visit the BTS website at:
https://www.brit-thoracic.org.uk/quality-improvement/lung-disease-registries/bts-ild-registry/

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# APPENDIX

## NHSE ILD Quality Dashboard Data (breakdowns by year)

This appendix includes a breakdown of figures by calendar year for six of the nine NHSE ILD Quality Dashboard items, which use IPF as a surrogate marker for the wider ILD service. Full details of the updated NHSE ILD Quality Dashboard items are included on the BTS website.

Data may be entered into the BTS ILD Registry prospectively and retrospectively, so the data in this appendix may vary in future data cuts (as records are added or amended). Some tables include responses of 'Not known/not recorded', which are typically excluded in the main text of this report.

Figures presented here include data from all participating sites, which area mixture of English specialist centres, English district general hospitals, and hospitals from the devolved nations.

**Item 1: IPF referrals discussed at ILD MDT**

*Percentage of new IPF referrals discussed at ILD MDT within 2 months of first assessment in the specialised service.*

This information is taken from Clinical Information question 2.8 *What was the outcome of the multi-disciplinary team meeting (MDT)?*

| | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| Awaiting MDT | 10 | 12 | 9 | 10 | 13 | 13 | 6 | 2 | | **75** |
| % | 7.1% | 4.1% | 2.5% | 3.2% | 3.3% | 2.7% | 1.6% | 1.3% | 0.0% | **2.9%** |
| Definite diagnosis of IPF | 66 | 127 | 158 | 142 | 179 | 227 | 167 | 60 | 24 | **1170** |
| % | 46.8% | 43.6% | 43.8% | 44.9% | 44.9% | 46.8% | 45.8% | 40.3% | 60.0% | **45.1%** |
| Likely diagnosis of IPF | 39 | 96 | 129 | 111 | 145 | 145 | 141 | 49 | 13 | **878** |
| % | 27.7% | 33.0% | 35.7% | 35.1% | 36.3% | 29.9% | 38.6% | 32.9% | 32.5% | **33.9%** |
| MDT not held | 21 | 25 | 25 | 13 | 9 | 10 | 6 | 6 | | **125** |
| % | 14.9% | 8.6% | 6.9% | 4.1% | 2.3% | 2.1% | 1.6% | 4.0% | 0.0% | **4.8%** |
| Working diagnosis of IPF | 5 | 31 | 40 | 40 | 53 | 90 | 45 | 32 | 3 | **345** |
| % | 3.5% | 10.7% | 11.1% | 12.7% | 13.3% | 18.6% | 12.3% | 21.5% | 7.5% | **13.3%** |
| **Total Count** | **141** | **291** | **361** | **316** | **399** | **485** | **365** | **149** | **40** | **2593** |

**Item 2: IPF patients offered or received ILD Specialist Nurse input**

*Percentage of new IPF referrals who were offered or received ILD Specialist Nurse input within 2 weeks of their first attendance in the service.*

This information is taken from Clinical Information question 2.23 *At the time of diagnosis, was the patient offered an interaction with an ILD specialist nurse? (This should not be more than two weeks after the first clinic visit).* This question was added to the Registry in December 2019.

| | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| Yes | 3 | 5 | 11 | 29 | 51 | 71 | 133 | 137 | 40 | 482 |
| % | 27.3% | 35.7% | 45.8% | 61.7% | 65.4% | 67.0% | 83.6% | 95.1% | 100.0% | 76.9% |
| No | 8 | 9 | 13 | 18 | 27 | 35 | 26 | 7 | | 145 |
| % | 72.7% | 64.3% | 54.2% | 38.3% | 34.6% | 33.0% | 16.4% | 4.9% | 0.0% | 23.1% |
| **Total Count** | **11** | **14** | **24** | **47** | **78** | **106** | **159** | **144** | **40** | **627** |

**Item 3: IPF patients assessed for pulmonary rehabilitation needs**
*Percentage of new IPF patients assessed for pulmonary rehabilitation needs.*

This information is taken from Clinical Information question 2.21 *Have you assessed if this patient is suitable to be referred to a pulmonary rehabilitation programme?*

Although data relating to pulmonary rehabilitation (PR) have been collected since the launch of the Registry, the PR question was updated in January 2017. No previous answers were mapped to the new question, as the content of the question had changed significantly. Therefore, all data included in this table were entered into the Registry from January 2017 onwards. All data from before 2017 were entered retrospectively, and these data should be interpreted with caution due to the reduced sample size.

| | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| Yes - assessed and referred | 6 | 10 | 18 | 41 | 183 | 274 | 198 | 55 | 29 | **818** |
| | 17.6% | 17.2% | 19.4% | 24.4% | 45.4% | 55.4% | 53.5% | 37.4% | 72.5% | **45.0%** |
| Yes - assessed but patient declined (does not wish it, no transport, etc.) | 4 | 2 | 11 | 15 | 32 | 33 | 21 | 10 | 2 | **131** |
| | 11.8% | 3.4% | 11.8% | 8.9% | 7.9% | 6.7% | 5.7% | 6.8% | 5.0% | **7.2%** |
| Yes - assessed but completed PR in the last 12 months | 1 | 2 | 2 | 11 | 23 | 31 | 11 | 4 | | **86** |
| | 2.9% | 3.4% | 2.2% | 6.5% | 5.7% | 6.3% | 3.0% | 2.7% | 0.0% | **4.7%** |
| Yes - assessed but not suitable (e.g. very poor mobility/very good fitness level already) | 6 | 17 | 16 | 31 | 64 | 74 | 85 | 45 | 8 | **347** |
| | 17.6% | 29.3% | 17.2% | 18.5% | 15.9% | 14.9% | 23.0% | 30.6% | 20.0% | **19.1%** |
| No - not assessed | 4 | 6 | 7 | 18 | 18 | 22 | 12 | 13 | | **101** |
| | 11.8% | 10.3% | 7.5% | 10.7% | 4.5% | 4.4% | 3.2% | 8.8% | 0.0% | **5.6%** |
| Not known | 13 | 21 | 39 | 52 | 83 | 61 | 43 | 20 | 1 | **335** |
| | 38.2% | 36.2% | 41.9% | 31.0% | 20.6% | 12.3% | 11.6% | 13.6% | 2.5% | **18.4%** |
| Total Count | **34** | **58** | **93** | **168** | **403** | **495** | **370** | **147** | **40** | **1818** |

**Item 4: IPF patients assessed for their palliative care needs**
*Percentage of new IPF patients assessed for palliative care needs.*

This information is taken from Clinical Information question 2.2 *Have you assessed and managed the palliative care needs of this patient at this clinic visit?*

| | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| Yes | 20 | 28 | 41 | 86 | 245 | 373 | 299 | 118 | 37 | **1254** |
| % | 58.8% | 47.5% | 39.8% | 50.6% | 61.1% | 76.0% | 80.6% | 78.7% | 92.5% | **68.6%** |
| No | 7 | 10 | 12 | 30 | 78 | 53 | 29 | 26 | 2 | **249** |
| % | 20.6% | 16.9% | 11.7% | 17.6% | 19.5% | 10.8% | 7.8% | 17.3% | 5.0% | **13.6%** |
| Not known | 7 | 21 | 50 | 54 | 78 | 65 | 43 | 6 | 1 | **326** |
| % | 20.6% | 35.6% | 48.5% | 31.8% | 19.5% | 13.2% | 11.6% | 4.0% | 2.5% | **17.8%** |
| **Total Count** | **34** | **59** | **103** | **170** | **401** | **491** | **371** | **150** | **40** | **1829** |

**Item 6: IPF patients assessed for their oxygen needs**

*Percentage of new IPF patients who have had their oxygen needs assessed (both long term and ambulatory oxygen).*

This information is taken from Clinical Information question 2.17a *Have you assessed the oxygen needs of this patient at this clinic visit?* This question was added to the Registry in January 2019. All data from before 2019 were added retrospectively.

| | 2013 | 2014 | 2015 | 2016 | 2017 | 2018 | 2019 | 2020 | 2021 | Total |
|---|---|---|---|---|---|---|---|---|---|---|
| Yes - assessed and referred (or already on oxygen) | 1 | 1 | 4 | 5 | 14 | 22 | 38 | 36 | 17 | **141** |
| | 11.1% | 6.3% | 14.8% | 10.4% | 18.4% | 19.5% | 25.9% | 27.1% | 43.6% | **23.0%** |
| Yes - assessed but patient does not require oxygen therapy at this time | 7 | 15 | 22 | 41 | 57 | 84 | 96 | 77 | 14 | **414** |
| | 77.8% | 93.8% | 81.5% | 85.4% | 75.0% | 74.3% | 65.3% | 57.9% | 35.9% | **67.5%** |
| Yes - assessed but patient declined (does not wish it, etc.) | 1 | | | 1 | 2 | 4 | 3 | 3 | 3 | **17** |
| | 11.1% | 0.0% | 0.0% | 2.1% | 2.6% | 3.5% | 2.0% | 2.3% | 7.7% | **2.8%** |
| Yes - assessed but not suitable (e.g. home environment unsafe for oxygen use) | | | | | 2 | | 1 | 3 | | **6** |
| | 0.0% | 0.0% | 0.0% | 0.0% | 2.6% | 0.0% | 0.7% | 2.3% | 0.0% | **1.0%** |
| Not – not assessed | | | 1 | 1 | 1 | 3 | 9 | 14 | 5 | **35** |
| | 0.0% | 0.0% | 3.7% | 2.1% | 1.3% | 2.7% | 6.1% | 10.5% | 12.8% | **5.7%** |
| **Total Count** | **9** | **16** | **27** | **48** | **76** | **113** | **147** | **133** | **39** | **613** |