BTS ILD Registry Annual Report 2020

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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 fifth 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 3,300 consenting people with the most common forms of interstitial lung disease: idiopathic pulmonary fibrosis and sarcoidosis. The data were collected over the last seven years from 73 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. Data leads to understanding, which leads to benefit.

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.

The BTS Interstitial Lung Disease Registry is relatively new (in comparison to similar registries in other countries) but is growing rapidly, and we are making refinements to ensure it meets its objectives. We continue to modify the consent and data entry processes to maximise benefits, reduce clinician burden, improve the quality of data and facilitate research and audit. We provide lay summaries to accompany this report, making the information accessible to patients looking to understand more about their condition (these lay summaries are available at https://www.brit-thoracic.org.uk/quality-improvement/lung-disease-registries/).

There have been a number of developments since the previous report, including:

- **Modification of the data fields** – a review of the Registry dataset was conducted, with the updated dataset launched in late 2019. Questions which could not be processed meaningfully were removed, such as occupation, bronchoalveolar differential cell count and echocardiography. Other questions were streamlined, including those regarding comorbidities and family history. A small number of new questions were added, including weight, BMI and place of death.
- **Revision of data validation criteria** – the data entry limits have been updated, which will reduce erroneous data entry. Lung function parameters and demographic details now have appropriate inbuilt checks to ensure implausible values cannot be entered.
- **Electronic consent** – approval has been granted to modify the consent process to permit eConsent (i.e. consent may now be obtained by electronic means).

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

The BTS ILD Registry continues to be a valuable resource which helps us understand two key elements about patient care: epidemiology and treatment. Previous reports have covered these elements in detail.

As this Registry is now in its eighth year another dimension is becoming increasingly apparent; the ability to analyse longitudinal data. This is a powerful tool which can be used to identify changes in practice over time, without which we would not be able to recognise improvements in care or quantify changes in the clinical landscape.

Since January researchers have also had the opportunity to apply to access anonymised Registry data. The UK IPF Registry in particular is already demonstrating the importance of facilitating research, and for the first time Registry data will be used in a large clinical trial (the TIPAL study).

Work is underway to make the BTS ILD Registry more user-friendly, and permission has already been received to allow the submission of electronic consent. Being responsive to the needs of clinicians – particularly in these challenging times – is essential to ensuring the Registry can continue to be used to improve care both locally and across the UK.

Professor Jonathan Bennett
Chair, BTS Board of Trustees

BTS Lung Disease Registry Steering Group Membership 2020:

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 Lisa Nicol, Consultant Respiratory Physician
Dr Katherine Spinks, Consultant Respiratory Physician

Mrs Sheila Edwards, BTS Chief Executive
Miss Sally Welham, BTS Deputy 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: John Bennett, a patient with IPF who sadly passed away in 2020. This picture is used with the kind permission of his wife, Dee. Courtesy of 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 2020, at a time when 67 sites across 56 Trusts/Health Boards had obtained approval to participate. The current full list of 73 participating sites is given on page 31.

Overall the BTS ILD Registry includes over 3,300 patient records (2,797 IPF records and 547 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 2,797 patient demographic records, not all questions are fully completed by centres for every patient. For example, family history was recorded for only 2,123 records and thus the denominator in this case would be 2,123.

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

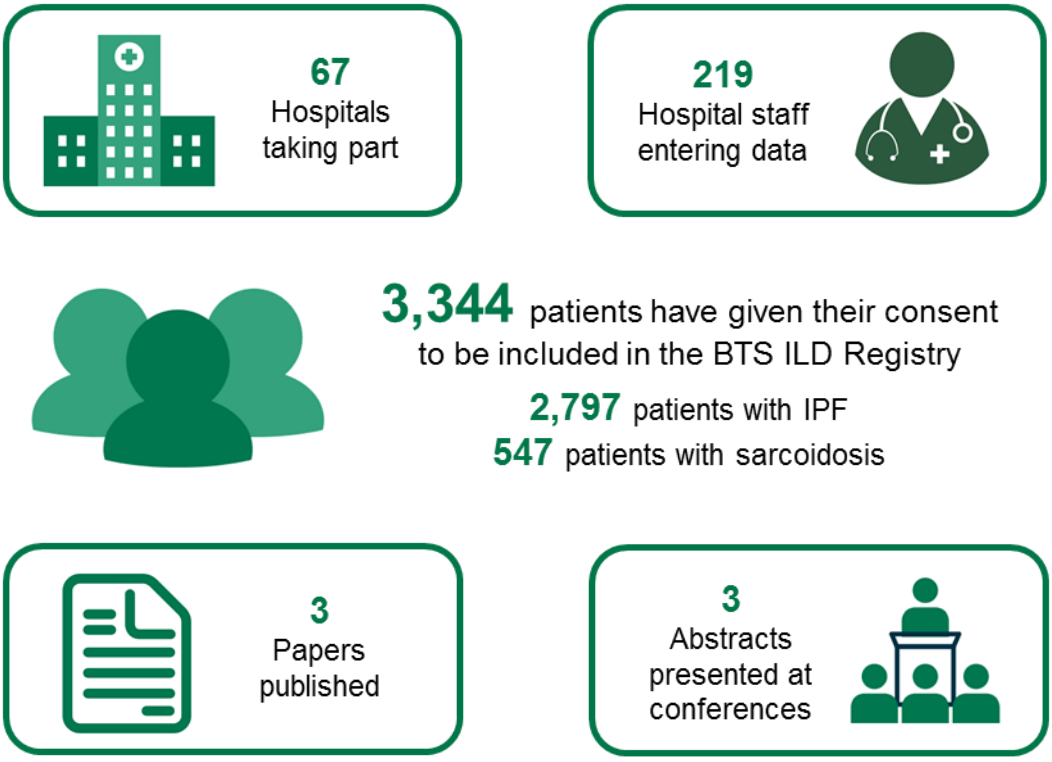
This report is based on data representing seven and a half years of hard work and dedication from those involved in the ILD Registry Steering Group and from the hundreds of Registry collaborators who have collected and entered data voluntarily.

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;
  - 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 and incurable lung disease. The symptom burden experienced by patients with IPF is high, with increasing symptoms of breathlessness, cough and fatigue. Some individuals also experience anxiety and depression. Survival remains poor and there are limited treatment options available.

Comorbidities are common and should be identified and addressed. Understanding the national burden of this condition is important for service provision and patient care. The British Lung Foundation's *Battle for Breath* report indicates that IPF resulted in 86,000 hospital bed days in the UK in 2011[2]. The UK IPF Registry, now it its 8th year, aims to report information that is relevant to patients with IPF, their carers, service commissioners and the wider community.

Unfortunately, there has been marked fall in the number of new entries to the database in 2020. Referrals to ILD centres for evaluation of IPF reduced as the respiratory community turned its attention to manage people with COVID-19 infection. In the first six months of 2020, 76 new patient records were entered into the Registry, which is a third of the expected number (a mean of 254 records were entered over the same period in 2018 and 2019). This limits the interpretation of recent trends. However, much of the data are presented as cumulative data since the Registry was established.

There have been a number of developments since the previous report, including:

- **Publication of research** – Dr Phil Molyneaux and colleagues (from the BTS ILD Registry Steering Group) investigated the consequences of using different algorithms for determining predicted lung function values, and the cost implications of changing to the Global Lung Index. The results were published at the 2020 ERS Congress[3]. Dr Helen Parfrey and colleagues have prepared an analysis of IPF Registry data from 2013-19, also on behalf of the BTS ILD Registry Steering Group (in press at the time this report was published[4]). Researchers from external organisations can also investigate outcomes from the Registry using the BTS Data Access Request Process (for more details visit https://www.brit-thoracic.org.uk/quality-improvement/bts-clinical-data-policy-and-data-access/).
- **Clinical trial involvement** – For the first time the UK IPF Registry is being used to collect data for use in a clinical trial. Approval has been given for data from participants of the randomised Treating Idiopathic Pulmonary Fibrosis with the Additional of Lansprazole (TIPAL) study to be collected via the UK IPF Registry where appropriate.
- **Reporting of longitudinal data** – In this report we present longitudinal data from the Registry for the first time, allowing us to highlight data trends over the last seven years.

There are several highlights from this report including:

- The length of time patients have symptoms before diagnosis remains concerningly long
- The decline in the number of lung biopsies taken before diagnosis
- The increasing age at presentation (with the exception of 2020)
- The stable, high burden of comorbidities
- The persistently low involvement of patients in clinical trials
- Increased palliative care assessments (with the exception of 2020)

This UK IPF Registry report presents data on the demographic, clinical/diagnostic criteria and follow-up records of patients, as well as highlighting how clinical activity maps to the NICE Quality Standard for IPF[1].

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 2020

At the end of June 2020, 67 centres had approval to participate in the UK IPF Registry, with 48 centres having contributed clinical data. As of December 2020, 73 centres have approval to participate (the full list of participating centres is given on page 31).

2,797 patient demographic records – an increase of 323 patients compared to October 2019

1,966 complete clinical/diagnosis records from the first clinic visit

1,533 Follow-up records representing 822 unique patients. These follow-up records represent 42% (822/1,966) 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

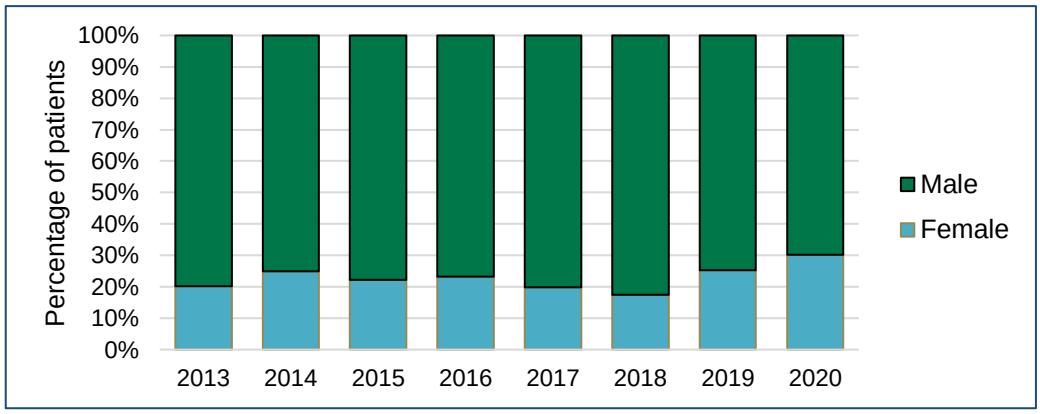


Figure 1: **Gender of patients over time**
The proportion of male and female patients has remained consistent over time, with a mean of 78% (2,105/2,701) males and 22% (596/2,701) females over the lifetime of the Registry. These data are in keeping with the known epidemiology of this disease.

Age

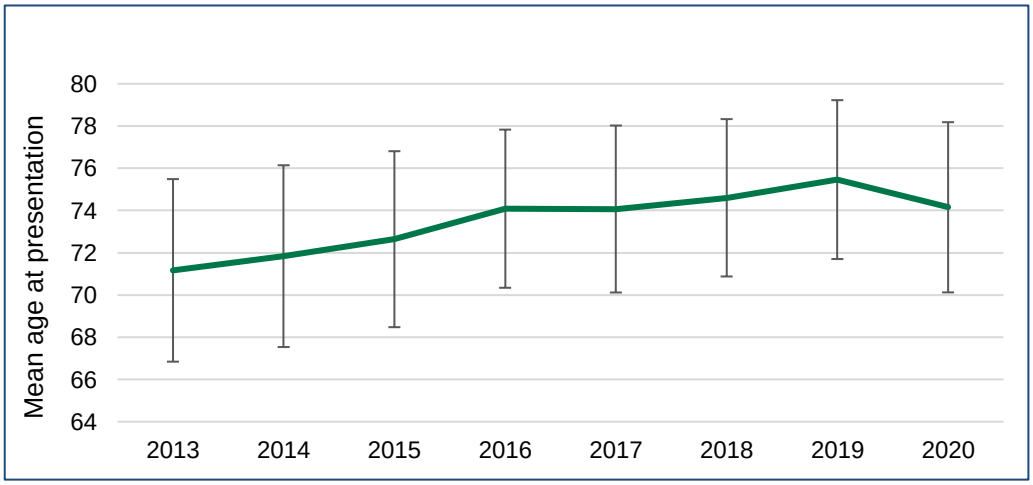


Figure 2: **Mean age at presentation over time**
The mean age of patients at presentation (shown here with error bars representing standard deviation) gradually increased over time from 71 years in 2013 to 76 years in 2019. In 2020 the mean age dropped to 74 years. However, the number of patient records in 2020 was very much lower than previous years.

Over the lifetime of the Registry the mean age was 74 years (± *8.1*), with 71% (1,882/2,653) aged 70 or over.

Comorbidities

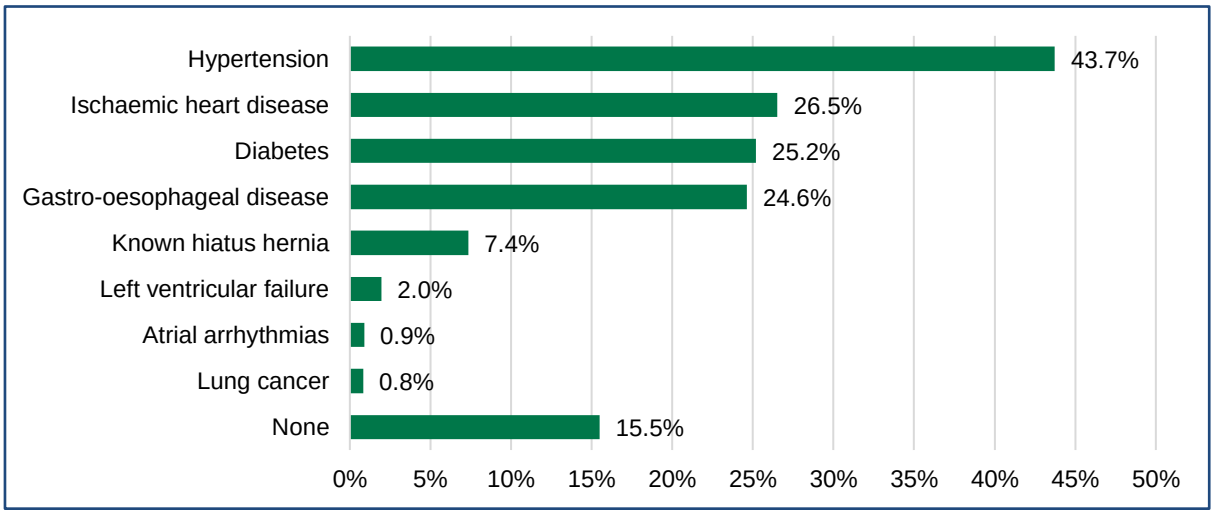


Figure 3: Comorbidities at presentation
Patients with IPF have a high burden of comorbidities at presentation, with 84.5% having at least one comorbidity. The nature and proportion of comorbidities remain stable over the lifetime of the Registry.

Smoking

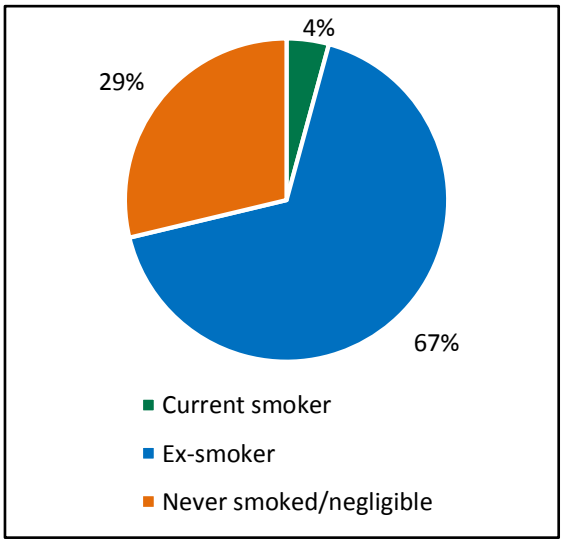


Figure 4: Smoking status at presentation
The proportion of patients who were smokers at presentation has remained consistent throughout the lifetime of the Registry. Overall, 4% (81/1,914) of patients were current smokers, 67% (1,283/1,914) ex-smokers and 29% (550/1,914) had never smoked or smoked a negligible amount (<5 pack years).

Relatives with IPF

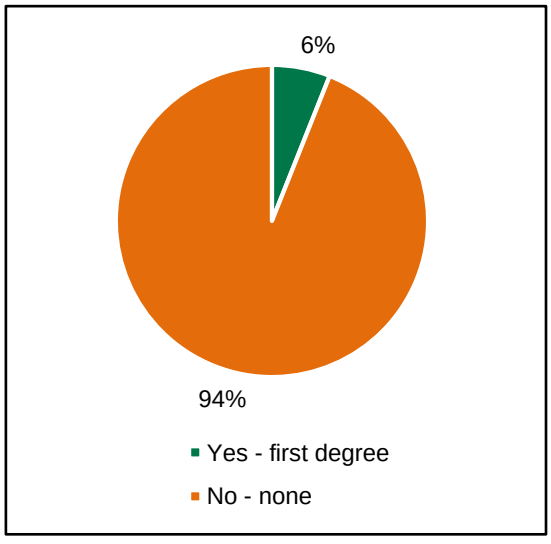


Figure 5: Known relatives with IPF
Where clinicians had recorded family history, 6% (128/2,123) of patients had a first degree relative known to have IPF and 94% (1,995/2,123) had no known relatives with IPF. These figures remained relatively consistent throughout the lifetime of the Registry.

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

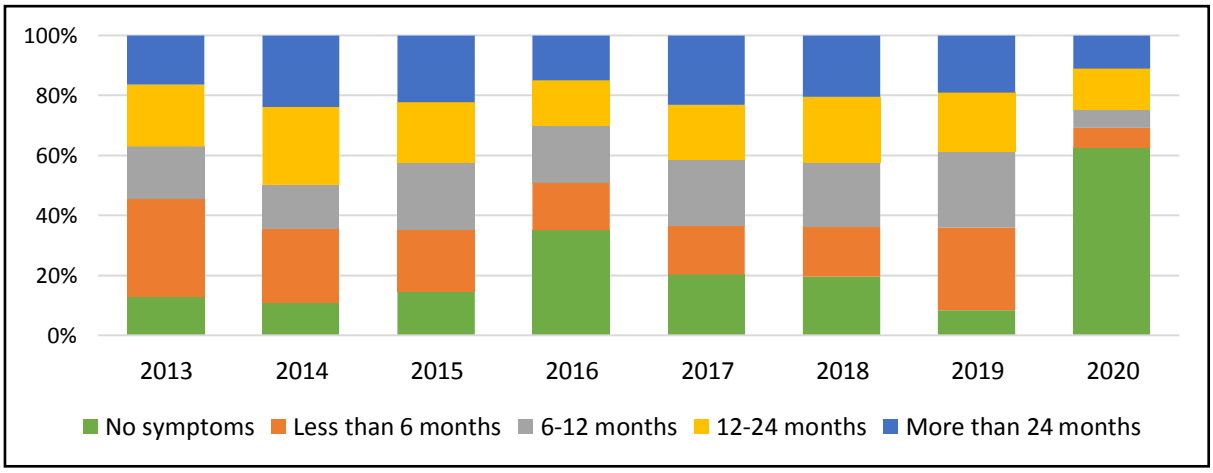


Figure 6: **Duration of IPF symptoms prior to presentation over time**

Unfortunately, patients often have symptoms for a considerable period prior to diagnosis. Since 2013, 41.5% (776/1,870) 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 be stable over time. Data from 2020 should be interpreted with caution, as only 6 months are included (to June) and the COVID-19 pandemic led to many fewer records being entered in this year.

Route of referral

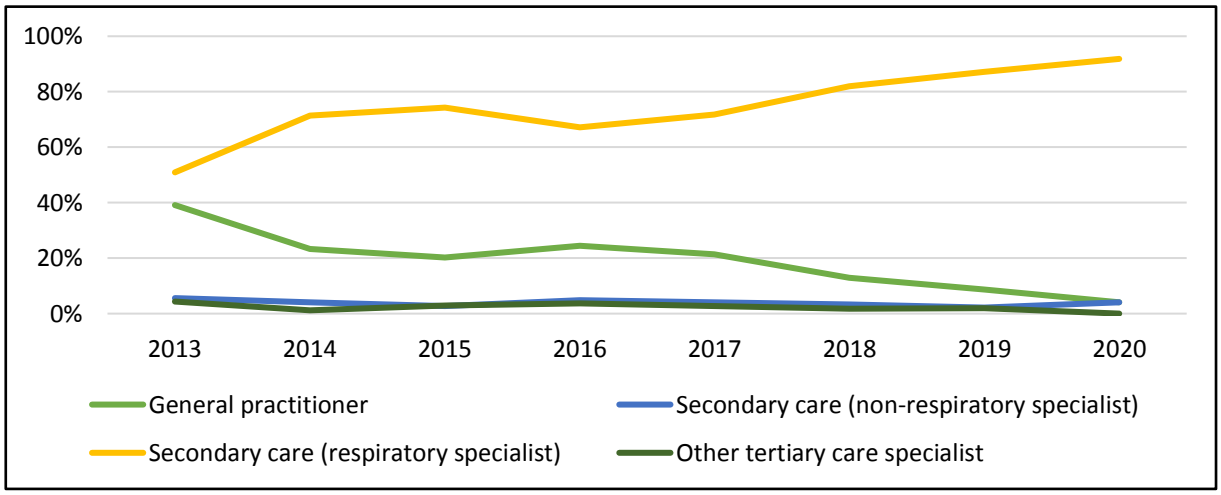


Figure 7: **Route of referral to clinic over time**

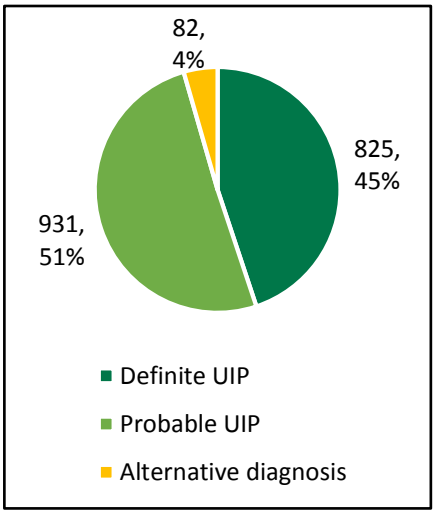
Throughout the lifetime of the UK IPF Registry the primary route of referral has been directly from a respiratory specialist in secondary care. Since 2013 there appears to have been a significant increase in referral from respiratory specialists in secondary care (from 51% in 2013 to 92% in 2020, although only 6 months of data are available for 2020) and a comparable reduction in referral from general practice (from 39% in 2013 to 4% in 2020). 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 13.1 weeks (13.6 for English specialist centres and 9.3 for other centres).

Mean waiting times from referral to multidisciplinary team (MDT) meetings were 10.8 weeks (10.7 and 11.1 for English specialist centres and other centres respectively).

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



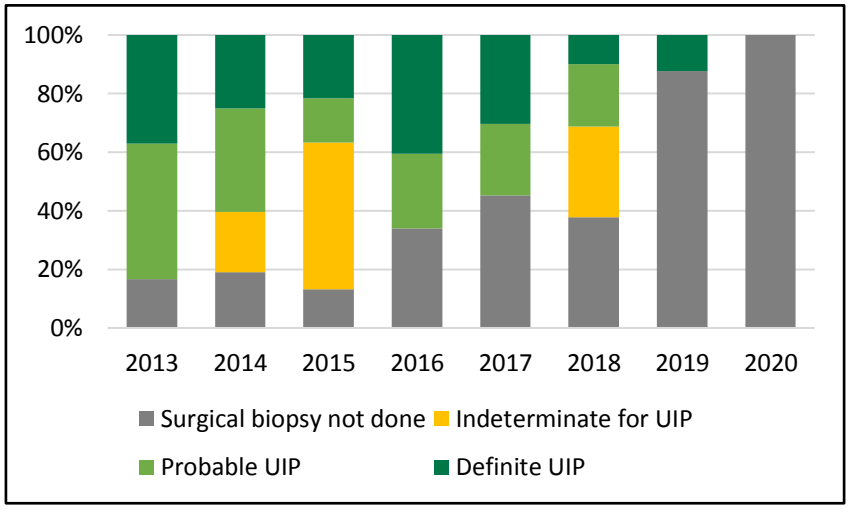


Figure 8 (left): **HRCT pattern at presentation, and**
Figure 9 (right): **Surgical biopsy data at presentation over time**
The majority of patients are diagnosed using high-resolution computed tomography (HRCT) scanning. Roughly equal proportions of these scans reported patterns in keeping with definite 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. The percentage of cases where a biopsy was conducted decreased over the last 7 years, especially since the diagnostic guidelines were updated in 2018. Data for 2020 represent only the first six months of the year (January to June).

MDT

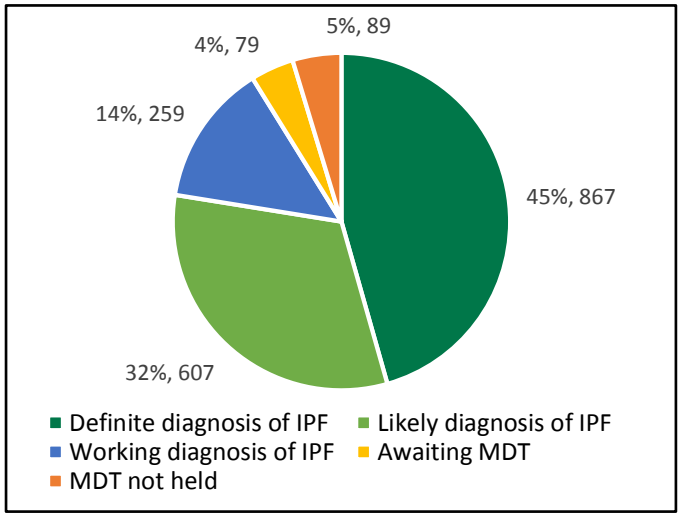


Figure 10: **Outcome of MDT**
The majority of cases are discussed at MDT (with 91% having been discussed at the first clinic visit and a further 4% due to be discussed at an upcoming MDT).

There appears to have been a steady fall in the number of cases not being discussed at MDT, reducing from 12.7% (17/134) of cases in 2013 to 0% of cases in 2019 (0/130).

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

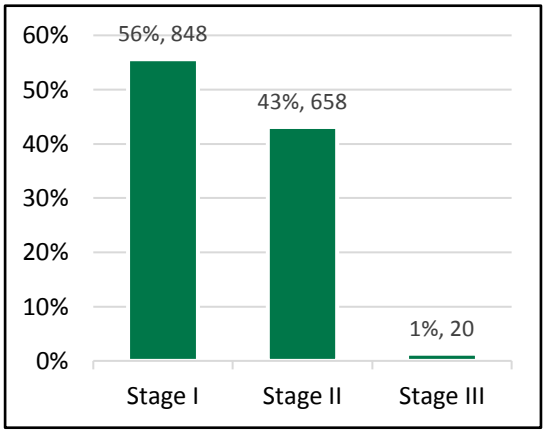


Figure 11: **GAP stage at presentation**
At first presentation the overwhelming majority of patients with IPF are categorised as having GAP Stage I or Stage II disease (99% of patients overall – 1,506/1,526). GAP staging is a marker of IPF disease severity, calculated using gender, age and lung function details[5].

Pulmonary hypertension

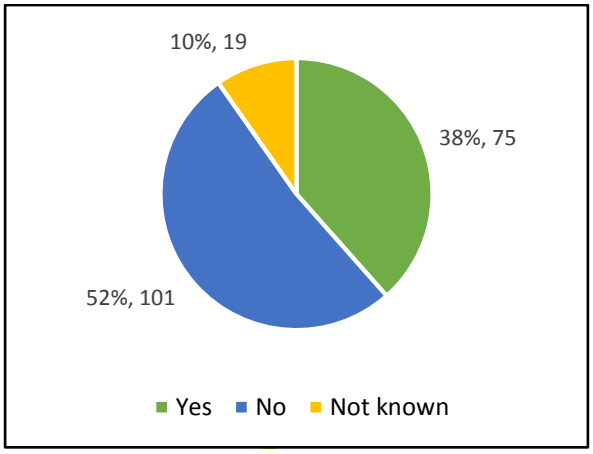
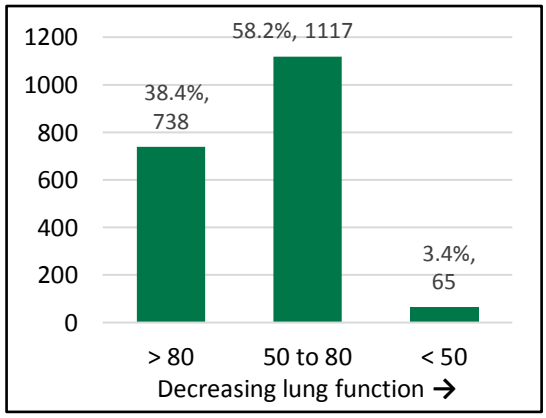


Figure 12: **History of pulmonary hypertension**
Overall 38% (75/195) of patients at presentation had previously been confirmed to have pulmonary hypertension or right heart strain, secondary to their lung disease, confirmed on echocardiogram or right heart catheterisation.

Lung Function (at presentation)



FVC Grouping	No. of patients	Mean DLCO % predicted	Standard Deviation
< 50%	38	36.7%	*15.1*
50 to 80%	879	45.9%	*15.7*
> 80%	602	53.0%	*15.9*

Figure 13 (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= 53%).

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 5% of patients on the UK IPF Registry have lower FVC values.

Drug treatment at presentation

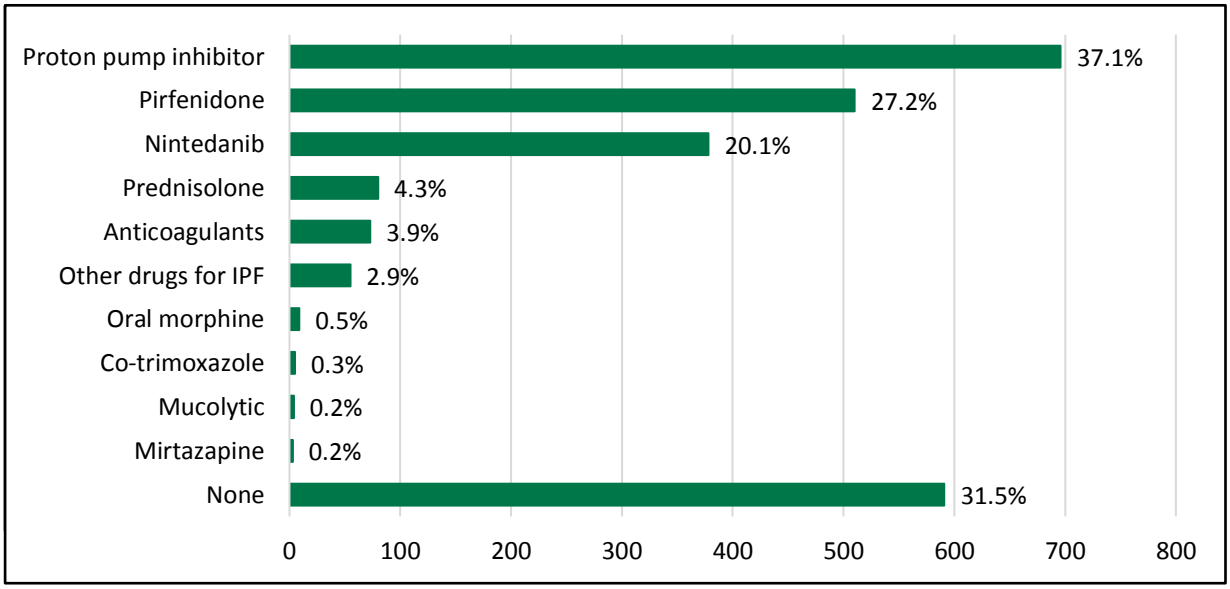


Figure 14: **Drug treatment at presentation (including treatment prescribed at first clinic visit)**
These data need to be interpreted carefully as they reflect drug use over the lifetime of the UK IPF Registry. Pirfenidone has been available through the NHS since 2013, whereas nintedanib has only been 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

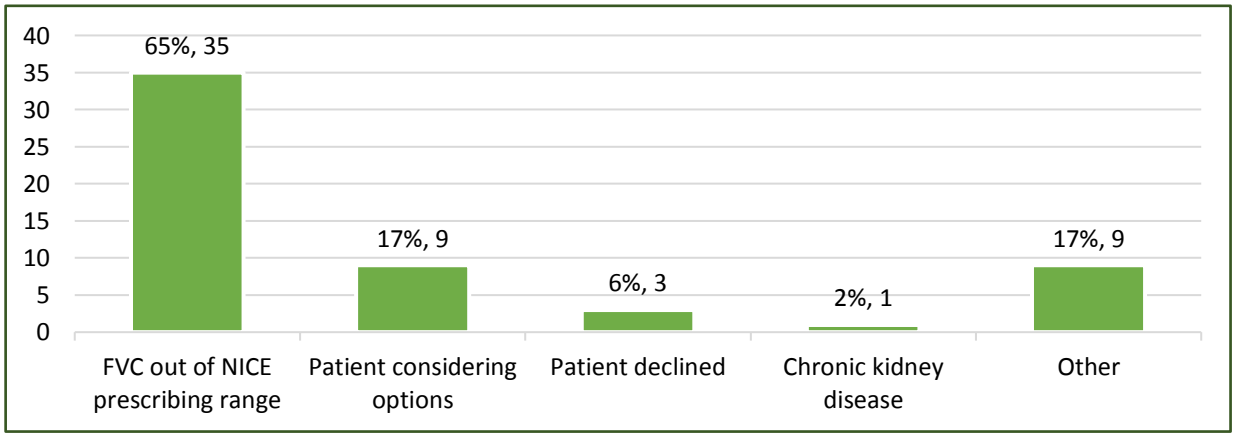


Figure 15: **Reason for not starting antifibrotic treatment at first clinic visit**
Since 2013, 53% of patients were not prescribed antifibrotic treatments at presentation. Suitability for drug treatment is not based purely on the FVC treatment criteria defined by NICE. Other factors, such as renal or 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 (65%) this was because the FVC was outside the range approved by NICE. For only 17% 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

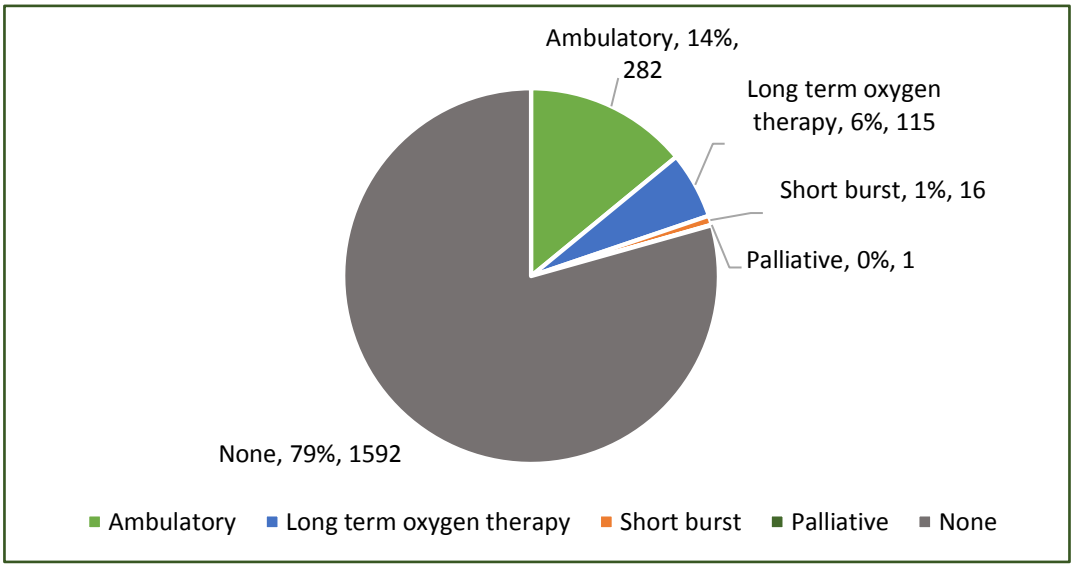


Figure 16: **Oxygen therapy at presentation**
At entry onto the Registry 92% (113/123) of patients had their oxygen needs assessed, with the majority (63%, 71/113) not requiring oxygen therapy at that time. One in six (17%, 331/1,923) patients were receiving or newly prescribed at least one form of oxygen therapy at their first clinic visit.

Lung Transplantation

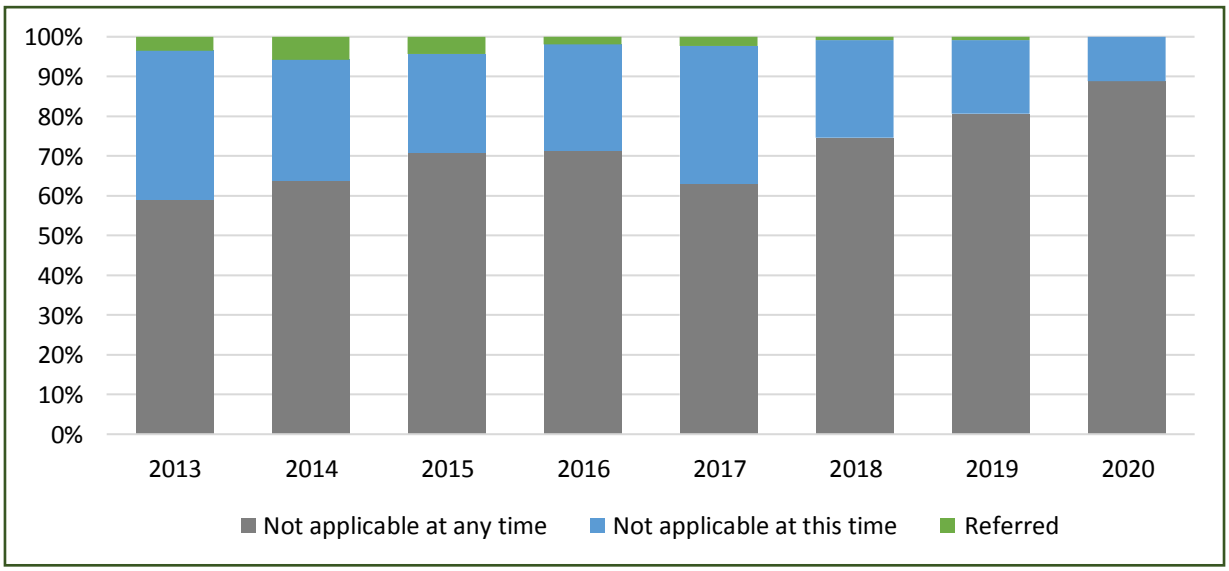


Figure 17: **Referral for lung transplantation at presentation over time**
Over the lifetime of the UK IPF Registry the proportion of patients deemed ineligible for lung transplantation 'at any time' at their first clinic visit appears to have increased gradually, rising from 59% in 2013 to 89% in 2020 (data for 2020 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 2, page 12).

Inclusion in Clinical Trials

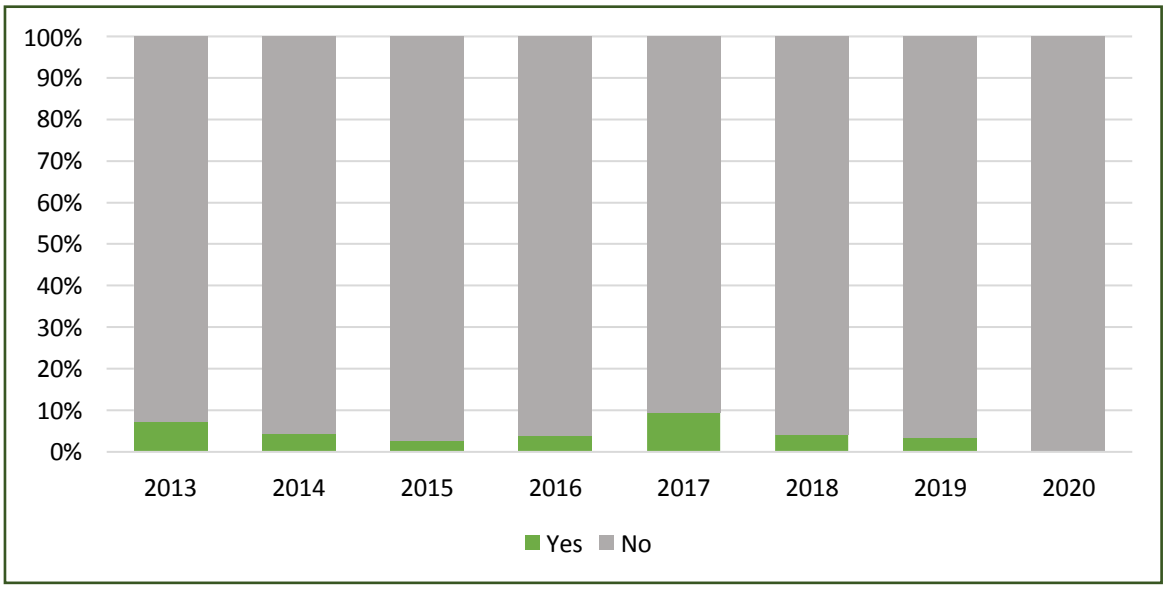
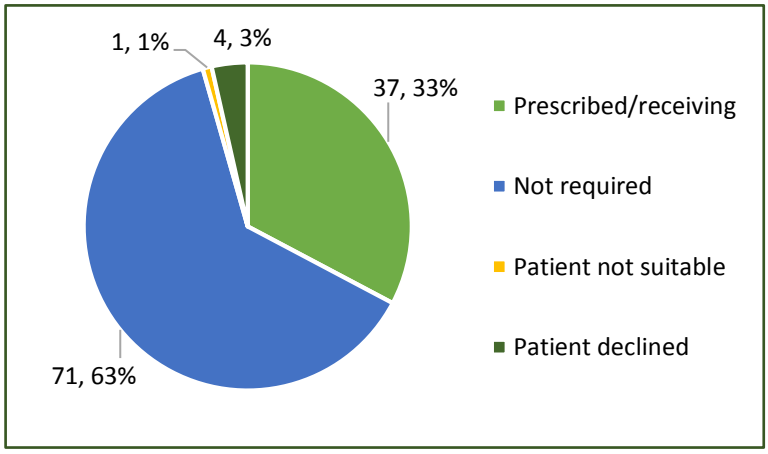
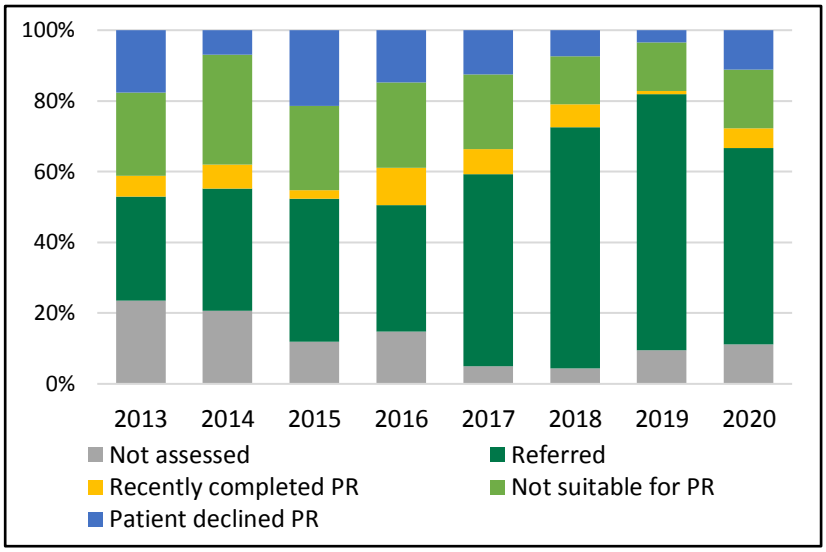


Figure 18: **Inclusion in clinical trials at presentation over time**
The proportion of patients recruited to clinical trials at the time of presentation has remained consistently low throughout the lifetime of the UK IPF Registry. Data from 2020 represent only the first six months of the year.

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: **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 91% of cases have already been discussed at MDT by the time of the first clinic visit and a further 4% due to be discussed at an upcoming MDT (see Figure 10, page 15).
Quality Statement 2: **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. At presentation 86% (111/129) of patients were offered the opportunity to interact with an ILD specialist nurse. Ideally this figure would be 100%.
Quality Statement 3: **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.**	 **Figure 19**: **Oxygen assessment at annual review** Clinicians are asked if they have addressed and managed the oxygen needs of the patient at follow-up clinic visits. Overall 92% (113/123) of follow-up visits included an oxygen needs assessment. Of those assessed the majority did not require oxygen therapy at the time (63%, 71/113). This question was added to the UK IPF Registry dataset in December 2019, therefore the number of responses is low.

Quality Statement 4: **Pulmonary rehabilitation (PR) programmes provide services that are designed specifically for IPF.**	 **Figure 20**: **Pulmonary rehabilitation (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, 92% (820/888) of patients had their PR needs assessed and 8% (68/888) did not. Of those assessed the majority (63%, 515/820) were referred for PR. There appears to have been an increase in the proportion of patients referred for PR over time, from 29% in 2013 to 72% in 2019 (dropping to 56% in 2020). 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). Additionally, data from 2020 represent the first six months only (to June). 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, 20% (161/820) were thought not to be suitable for referral, either due to poor mobility or already having a good fitness level.
Quality Statement 5: **People with IPF and their families and carers have access to services that meet their palliative care needs.**	At presentation 81% (701/868) patients were reported to have had their palliative care needs assessed. By palliative care we mean care intended to optimise quality of life and reduce suffering (for example, through symptom management). When completing the question regarding palliative care assessment, clinicians are advised that the patient's needs have still been assessed even if that assessment identifies no current need for palliative support.

PART 3 – The UK Sarcoidosis Registry

Sarcoidosis is a disease with an incidence of around 7 per 100,000 per year in the UK[6]. It is characterised by granulomatous inflammation in any organ, although the lungs are most commonly affected (over 90% of patients). Other commonly affected organs are the eyes and skin. It typically presents between 20 and 50 years of age, but can occur in those over 60. These patients are largely managed in outpatient services in secondary care, although the British Lung Foundation's *Battle for Breath* report indicates that sarcoidosis resulted in 9,000 hospital bed days in the UK in 2011[2].

There is significant heterogeneity in the disease, with variability in when patients present to clinicians, the volume of disease, the numbers of organs involved, disease course and response to pharmacological treatment. The UK Sarcoidosis Registry provides an opportunity to map and distil the variation in sarcoidosis across UK, ultimately leading to improvement in management and patient care. The Registry is one of a number of Quality Improvement initiatives designed and implemented through BTS, including the BTS Clinical Statement on Pulmonary Sarcoidosis (in press at the time this report was published)[7].

Since its inception, the UK Sarcoidosis Registry has amassed over 540 cases from across the UK. Data collection through a national registry provides a number of opportunities, such as the ability to collect data for the burden of disease on employment. It also highlights a challenge faced by clinicians: stretched resources making it extremely difficult to input information into voluntary initiatives such as the Registry. The report shows latest data which will form the basis of potential changes in the coming years.

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 2020

At the end of June 2020, 67 centres had approval to participate in the UK Sarcoidosis Registry, with 38 centres having contributed clinical data. As of December 2020, 73 centres have approval to participate (the full list of participating centres is given on page 31).

547 patient demographic records

390 complete clinical/diagnosis records from the first clinic visit

185 follow-up records representing 100 unique patients.

3.1 THE SARCOIDOSIS PATIENT COHORT

Patient demographic information is collected at the first clinic visit.

Gender

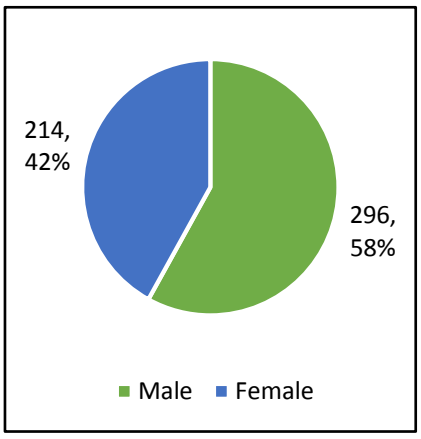


Figure 21: **Gender of patients**
The proportion of male (58%, 296/510) and female (42%, 214/510) patients has remained consistent over the lifetime of the UK Sarcoidosis Registry.

Age

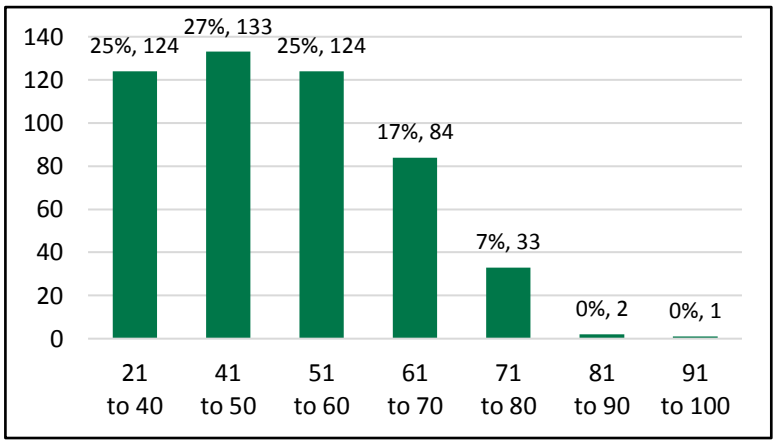


Figure 22: **Age at presentation**
Almost a quarter of patients (24%, 120/501) presented over the age of 60, with the mean age at presentation being 50 years, with a standard deviation of ± 13.4.

Ethnicity

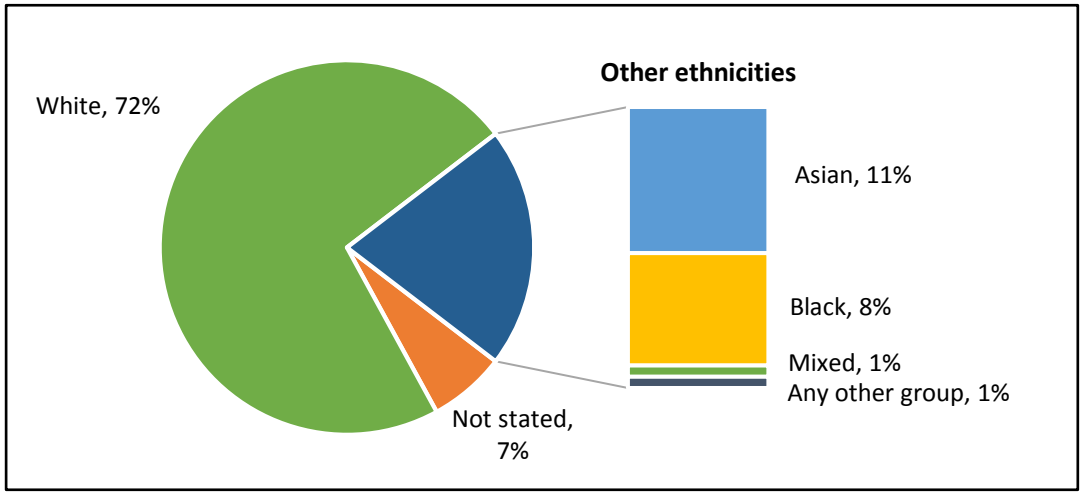


Figure 23: **Patients by ethnic group**
The majority of patients in the Registry were white (72%, 349/481). Although sarcoidosis in known to be more prevalent in black populations, only 8% (40/481) of patients were black. This figure likely reflects the populations from which Registry data were obtained, 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[8].

Smoking status

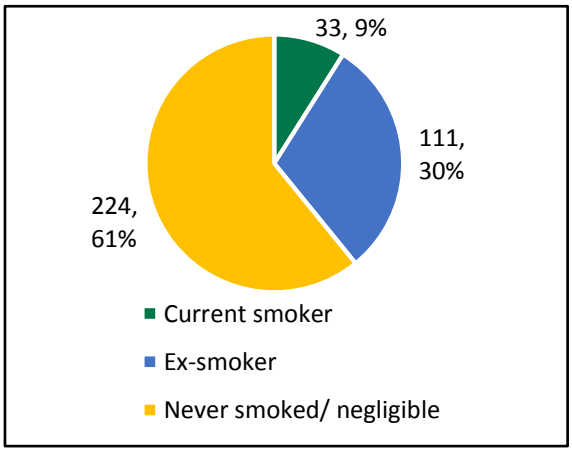


Figure 24: **Smoking status at presentation**
Overall 39% (144/368) of patients were either smokers or ex-smokers at presentation (patients were counted as ex-smokers if they quit more than three months before their first clinic visit).

61% (224/368) 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 (65%, 184/283) had no reported comorbidity at the time of their current presentation

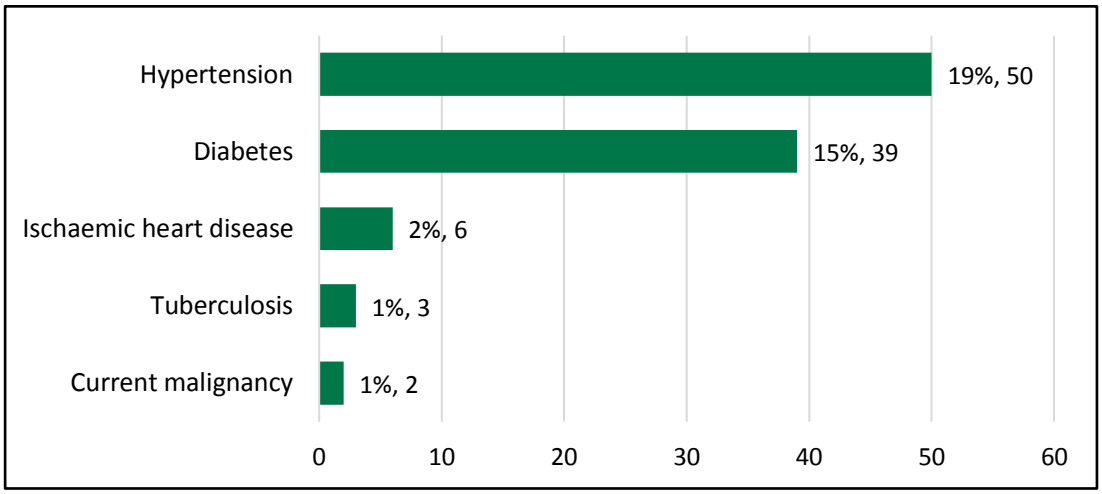


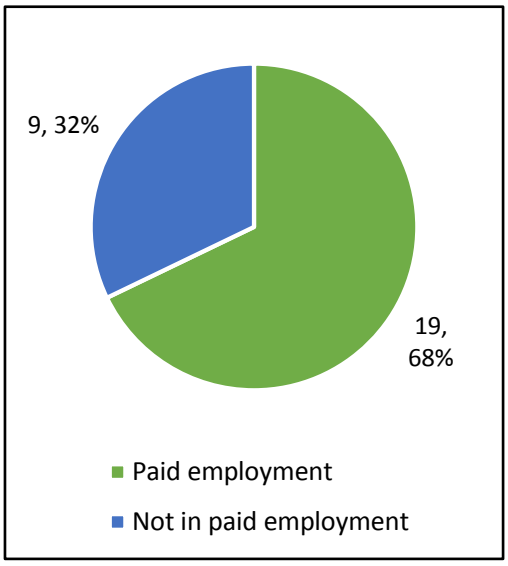
Figure 25: **Patient comorbidities at presentation**
The most commonly reported comorbidities were systemic hypertension (one in five patients – 20%, 57/283) and diabetes (15%, 43/283). These conditions are highly prevalent in the general population. Where at least one comorbidity was recorded patients 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% (14/398) 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 26: **Employment status at presentation**
Where employment status at presentation was known, over two thirds (68%, 19/28) of patients were in paid employment. Of those who were not in paid employment the majority (78%) were retired.

3.2 DIAGNOSING SARCOIDOSIS

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

Referral to clinic

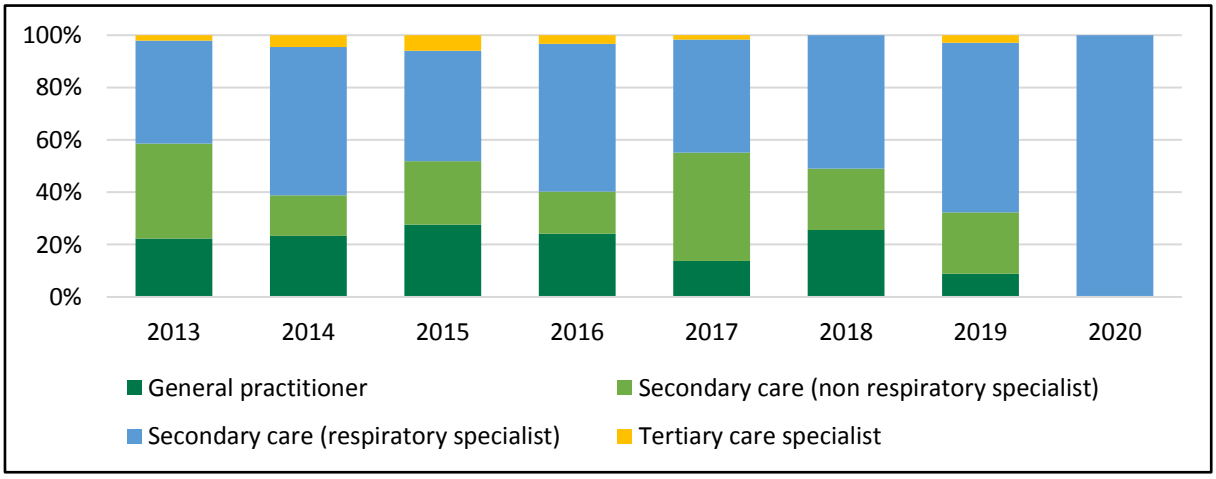


Figure 27: **Route of referral to clinic over time**

Overall, 49% (240/489) of patients were referred from respiratory physicians in secondary care. Referrals from general practice have remained low at 23% overall (110/489), 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 40% (149/373) of cases being diagnosed incidentally. Data from 2020 should be interpreted with caution, as they represent only the first six months of the year (to the end of June) and the COVID-19 pandemic led to many fewer records being entered.

Symptoms at first clinic visit

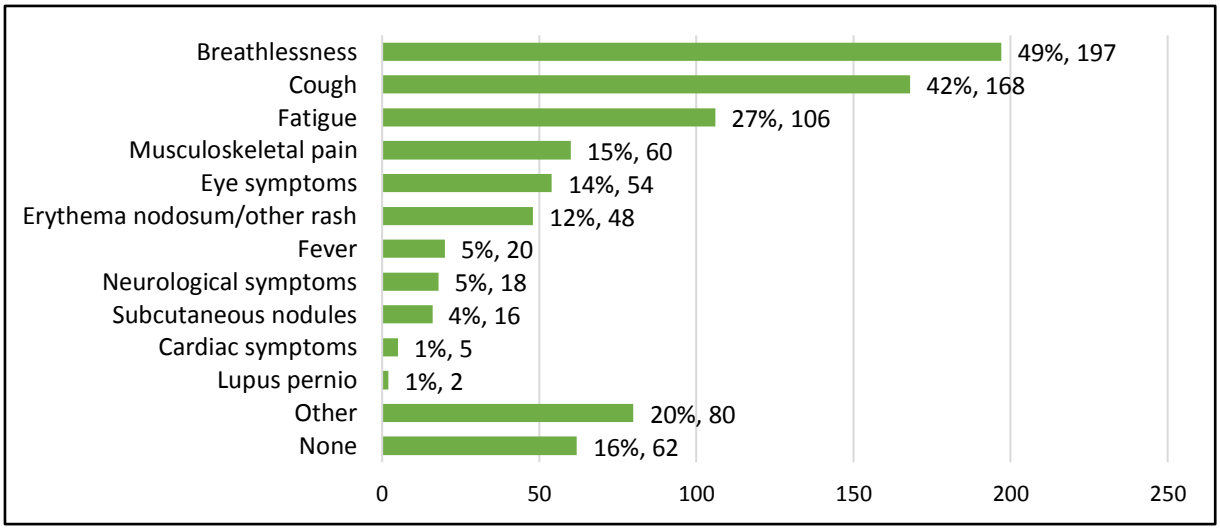


Figure 28: **Symptoms reported at first clinic visit**

The most common symptoms were breathlessness (49%), cough (43%) and fatigue (27%). Musculoskeletal pain (15%), eye symptoms (14%) and skin rashes (12%) 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 96% (292/304) 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.

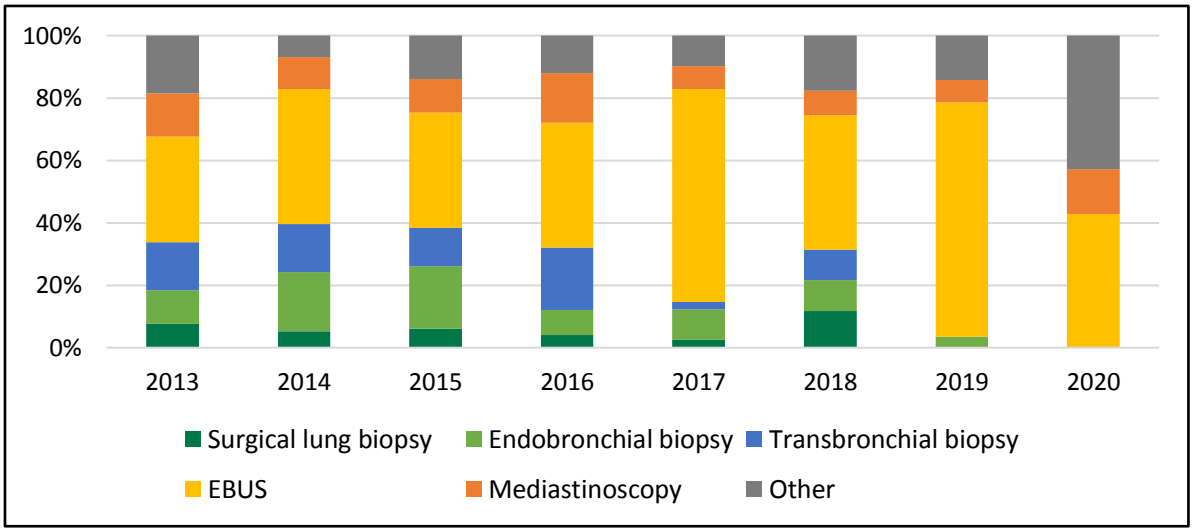


Figure 29: **Biopsy techniques used at presentation over time**
A number of different biopsy techniques were employed. Endobronchial ultrasound (EBUS) was consistently the most popular technique, accounting for 57% (165/292) of all biopsies conducted. Data from 2020 should be interpreted with caution, as they represent only the first six months of the year.

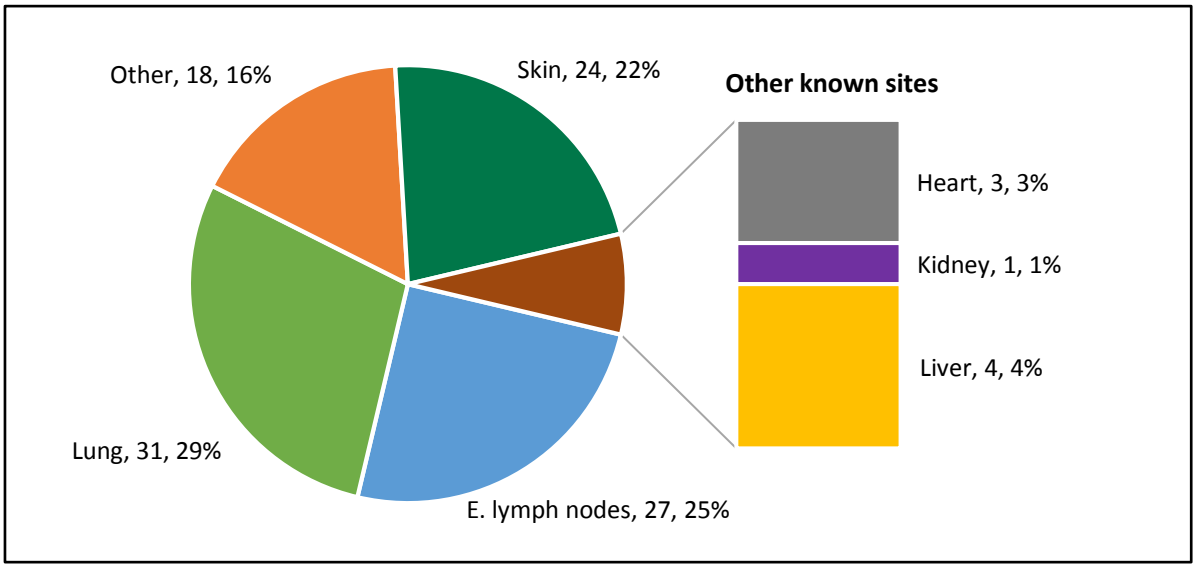


Figure 30: **Site(s) from which histology obtained at presentation**
The most common biopsy sites were lung (29%, 31/108), extra-thoracic lymph nodes (25%, 27/108) and skin (22%, 24/108). The variety of biopsy sites investigated is indicative of the multisystem involvement commonly observed in sarcoidosis.

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

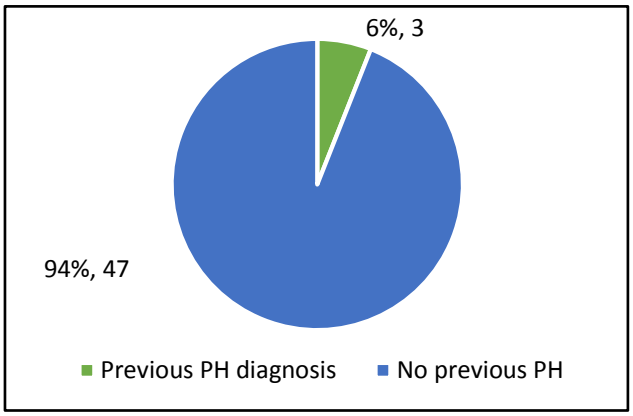


Figure 31: **History of pulmonary hypertension (PH)**

At presentation 6% (3/50) 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

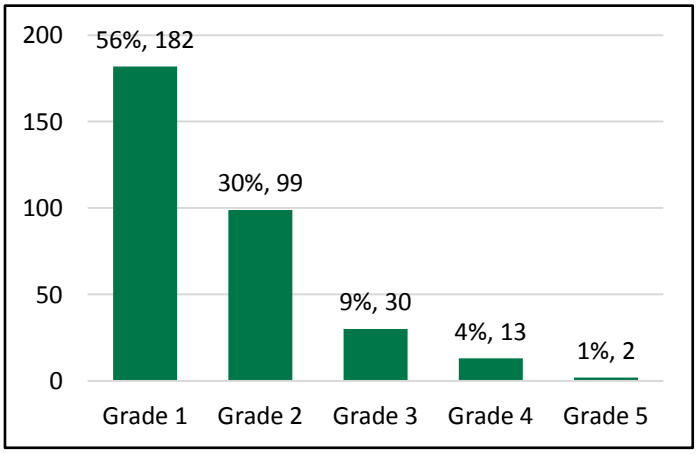


Figure 32: **Breathlessness at presentation**

The majority of patients (86%, 281/326) 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

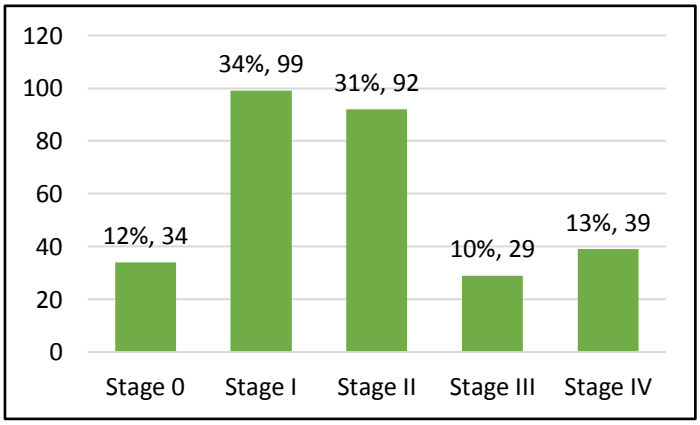


Figure 33: **Chest radiograph at presentation**

At presentation the majority of patients were observed to have either Stage I with lymphadenopathy only (34%, 99/293) or Stage II lymphadenopathy and parenchymal involvement (31%, 92/293) disease according to the Siltzbach sarcoidosis classification system.

Blood tests

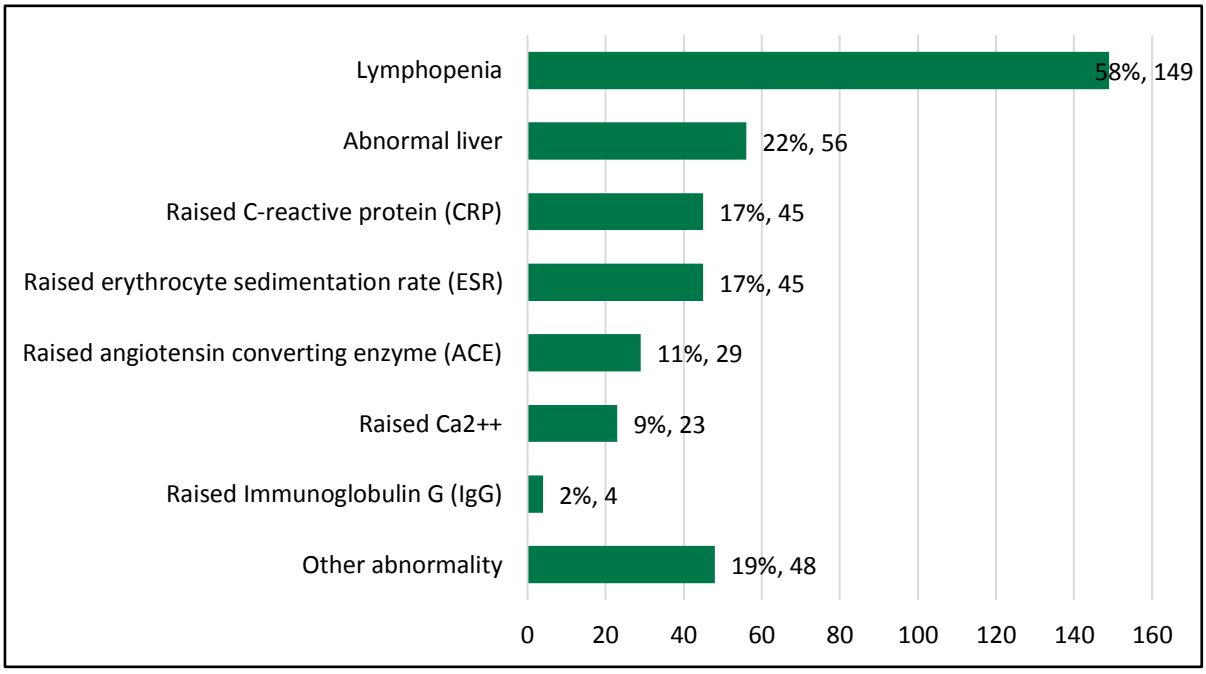


Figure 34: **Blood test abnormalities recorded at presentation**
The most common abnormality recorded in blood tests at presentation was lymphopenia, identified in 57% (149/263) of patients. Raised Immunoglobulin G (IgG) was only identified in 2% (4/263) of patients.

HRCT pattern

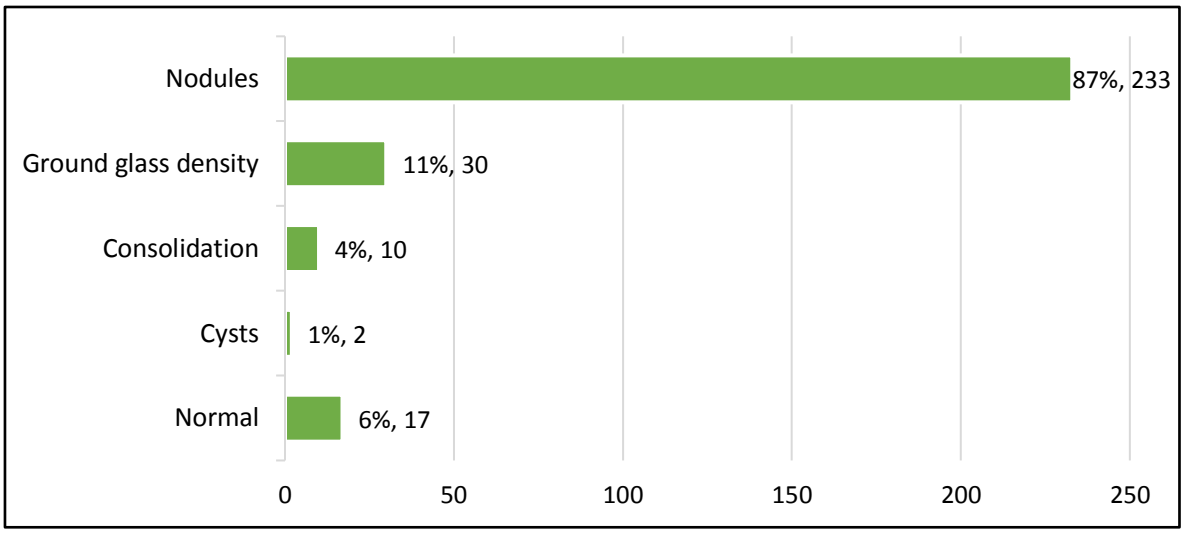


Figure 35: **Parenchymal abnormalities identified on HRCT at presentation**
The most common parenchymal abnormality identified on HRCT imaging at presentation was nodules, found in approximately seven out of every eight cases (87%, 233/269).

Current drug treatment

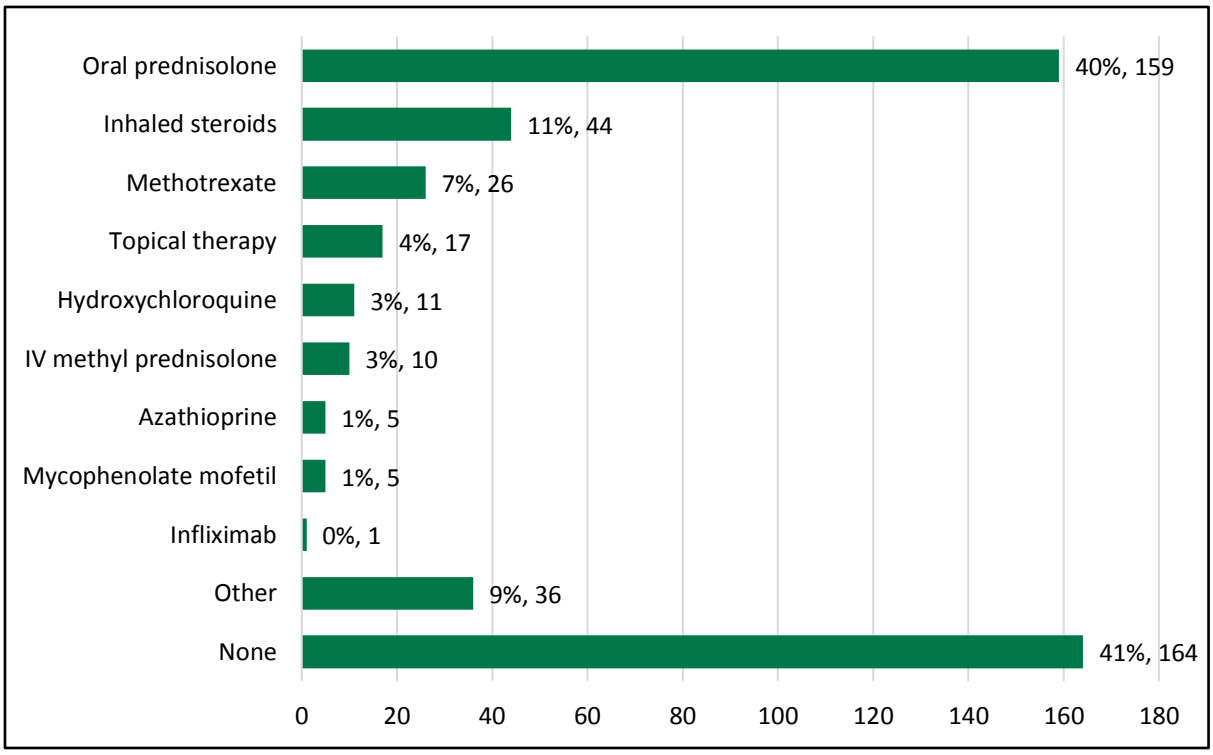


Figure 36: **Drug treatment at presentation**
The majority of patients were either not started on treatment (41%, 164/396) or managed with systemic corticosteroids (48%, 190/396). A number of aternative agents were used, with none used in more than 11% of cases. This broadly reflects previous BTS guidance on the management of sarcoidosis[9].

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 27% (8/30) of patients were referred or signposted to other services at the time of presentation. Of those who were, 38% (3/8) were given details for both patient support groups and helplines.

Inclusion in clinical trials

At presentation only 1% (5/373) 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
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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