British Thoracic Society

UK IPF and UK Sarcoidosis Registries

Ten Year Report | 2013-2023

"BTS is leading the way in harmonising the capture of valuable demographic and clinical information on IPF and sarcoidosis patients onto a national database to enable better understanding of these chronic conditions. The data will establish indicators of best practice and help us to enhance the quality of care for patients with IPF and sarcoidosis across the country."

Professor Monica Spiteri,
Speaking in 2014 as the then Chair of the BTS Lung Disease Registry Steering Committee

"BTS is proud that the Registry allows all UK participating sites to better achieve this and, in England, to complete ILD Quality Dashboard returns. It is also delighted that the Registry is a conduit to increased clinical trial participation, which will also lead to better care."

Dr Paul Walker,
Speaking in 2023 as the Chair of BTS

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British Thoracic Society
Registered Office: 17 Doughty Street / London WC1N 2PL
T: + 44 (0) 20 7831 8778 F: +44 (0) 20 7831 8766
bts@brit-thoracic.org.uk • www.brit-thoracic.org.uk

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FOREWORD: DATA SHARED TO IMPROVE CARE

Welcome to the 7th annual report from the BTS Interstitial Lung Disease Registry, which includes data from the UK IPF Registry and the UK Sarcoidosis Registry.

The Registry has completed its 10th year and therefore this report is a celebration of the last decade of work voluntarily undertaken by numerous clinicians, data managers, administrators and researchers, supported by a rolling BTS Steering Group and British Thoracic Society Head Office. More importantly, the Registry is supported by Action for Pulmonary Fibrosis and SarcoidosisUK, the UK National Interstitial Lung Disease patient charities, and more than 6000 patients who have agreed to permit their data to be included.

This report provides data on the current status of IPF and sarcoidosis in the UK in terms of demographics, comorbidities, referral and diagnostic pathways, treatments, and outcomes in terms of lung function and survival. However, it also provides an opportunity to reflect on the changes over the last 10 years and to note key differences particularly in terms of diagnosis. In keeping with other reports, this report provides data on the Idiopathic Pulmonary Fibrosis Quality Statements, and it is heartening to see steady improvement in the percentage of people meeting these standards over the years, which for some standards is substantial. External peer reviewed reports generated from the data are highlighted in this report and there are more in preparation.

There has been a steady increase in contribution to the Registry in terms of patients consenting that their data may be reported and also the numbers of centres entering data. Although much of the data is provided by specialist centres, 87 individual sites have enrolled to participate in this venture, representing sites from throughout the devolved nations and ensuring generalisability in the data. With data from 5,052 individuals with IPF and 937 with sarcoidosis, the Registry is one of the world's largest of its type.

There have been modifications to the dataset over time to align with current needs. However, great care has been taken to ensure that data can be mapped from one dataset to another where possible to permit longitudinal review. There have also been changes in terms of mandatory requirement for specialist centres, improvements in the consent process to facilitate easier consent without reducing its rigor, revision of quality dashboards and validation of the data. As mentioned in detail in the report, the most significant recent change is the amalgamation of the two registries with inclusion of all people with fibrotic interstitial lung disease.

However, the fundamental values of the Registry hold firm: to improve the quality of care for people with interstitial lung disease and thereby improving outcomes. As evidenced by the data, and also the anecdotal quotes from stakeholders, the Registry has already had an impact on patient lives. Clinicians also benefit from benchmarking and researchers from data access opportunities; there is a role for anyone involved with ILD.

I hope you find this report useful and you will continue to support the Registry in any way you can so we can continue on its journey of improving the lives of people with this dreadful condition.

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

Starting on a journey always involves uncertainty which can excite, fascinate and cause anxiety, sometimes in equal measure. Eleven years ago, no one involved knew how the Registry would develop and what it would become. The vision, expressed by Monica Spiteri in 2014, was to "enhance the quality of care for patients with IPF and sarcoidosis across the country". I hope that all individuals involved are proud of the first 10 years.

This report represents the end of one journey and the start of another; it includes data from the IPF Registry and Sarcoid Registries and, in future, reports will be for the combined ILD Registry. Registries contain real life data which can be messy, unpredictable and at times lack clarity. But we practice real life medicine so this is the world in which we operate and of which we are familiar. This report highlights what happens to people with IPF and sarcoid including time to diagnosis, investigation, MDT involvement, access to services, treatment and outcomes. This information is really important and the Registry has already been used to disseminate insight and analysis, via abstract and publication, and we hope this continues and expands over the next 10 years.

Healthcare metrics are vital to benchmark services, drive quality improvement and improve the care we deliver. BTS is proud that the Registry allows all UK participating sites to better achieve this and, in England, to complete ILD Quality Dashboard returns. It is also delighted that the Registry is a conduit to increased clinical trial participation, which will also lead to better care.

Ultimately, the Registry exists only by the engagement, hard work and dedication of patients and carers, healthcare professionals who submit data, the BTS team who administer the database and individuals, especially Registry Steering Group members, who dedicate their time. The Society and I are grateful to all, from launch to now, without whom the Registry would not be the success it is. Certainly, the next journey is a very exciting one.

Dr Paul Walker
Chair, BTS Board of Trustees

BTS Lung Disease Registry Steering Group Membership 2023:

Professor Andrew Wilson, Chair
Mrs Sarah Agnew, Nurse Representative
Mr Howard Almond, Patient Representative
Mr Leo Casimo, SarcoidosisUK
Dr Ahmed Fahim, Consultant Respiratory Physician
Dr Sophie Fletcher, Consultant Respiratory Physician
Dr Sarah Haney, Consultant Respiratory Physician
Professor Ling-Pei Ho, Consultant Respiratory Physician
Dr Clare Hodkinson, Action for Pulmonary Fibrosis
Dr Paul Minnis, Consultant Respiratory Physician
Dr Evelyn Palmer, Trainee Respiratory Physician
Dr Iain Stewart, Volunteer member

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

BTS would like to thank all those who have previously been involved in the Lung Disease Registry from its inception in 2013 to the launch of the new, expanded Registry in 2023.

- Our former chairs: Professor Martyn Partridge (2011 – 2013), Professor Monica Spiteri (2013 – 2017) and Dr Lisa Spencer (2018 – 2020).
- All those who previously served on the steering group, generously volunteering their time and expertise.
- The clinicians, nursing staff and administrative staff who diligently consented patients, collated information and submitted data to the Registry over the last decade.
- All the patients who have kindly consented to take part in the Registry – none of this work would be possible without their support.

Cover photograph: With thanks to Maggie Bartlett, a patient living with interstitial lung disease, and her husband and carer, Ian Maries. Image courtesy of Action for Pulmonary Fibrosis.

INTRODUCTION

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

These registries were developed with the aim of improving standards of care for patients with IPF and sarcoidosis, including through enabling and facilitating research to improve understanding of the epidemiology and progression of these diseases.

During 2022, as BTS approached ten years since the launch of these registries, a review was undertaken to determine how the Lung Disease Registry Programme could better support patients, clinicians and researchers going forward. Consequently, a new BTS UK ILD Registry was formed.

This new Registry was launched in February 2023, and it includes the existing data previously collected for IPF and sarcoidosis. The BTS UK ILD Registry builds on the work of the registries for IPF and sarcoidosis, expanding to collect information about all fibrosing ILDs (as well as non-fibrosing sarcoidosis). This expansion of the Registry will support a greater understanding of how cases of fibrosing ILD are treated nationwide.

So while this report will be the final report for the UK IPF and UK Sarcoidosis Registries, the data here have been incorporated into the new BTS UK ILD Registry and will be included in future reports (as well as still being available for researchers to apply to access).

Collating data from a larger group of people across time helps to improve understanding of the broader ILD landscape. The creation of a more substantial knowledge base gives a means of improving epidemiology and subsequently patient care. Patients suffering from, and clinicians treating, ILDs that have a lower prevalence see particular benefit from this collation of national data. This Registry helps to build greater equality in care by providing insight into rarer illnesses that otherwise would struggle to receive focus.

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 19th February 2023, at a time when 81 sites across 65 Trusts/Health Boards had obtained approval to participate. This current full list of 87 participating sites is given on page 35.

Overall, the BTS ILD Registry includes just shy of 6,000 patient records: 5,052 IPF records and 937 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 (22/EE/0235) has been granted by the NRES Committee East of England. It was first granted in October 2012, then renewed in October 2017 and again in November 2022. 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.

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 – Ten Years of the BTS ILD Registry

This report is based on data representing ten 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.

1.1 IMPACT OF THE BTS ILD REGISTRY

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 drive 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.

There have been a number of abstracts and manuscripts published in peer reviewed respiratory journals since the conception of BTS IPF and Sarcoidosis Registries in 2013 (Table 1, overleaf). These publications have described important aspects of IPF and sarcoidosis care in the UK and highlighted limitations on the NICE criteria in terms of Forced Vital Capacity (FVC) on eligibility of anti-fibrotic therapy with regards to reference equations used for FVC calculation (Phil Molyneaux *et al.*, 2020[2]).

We expect significant further publications in the near future addressing important aspects of ILD. This is especially true with the recent amalgamation of the IPF and sarcoidosis registries, and the expansion to include all ILDs with evidence of fibrosis. This increase in scope will provide invaluable data on the natural history of these diseases, as well as helping to tackle many key un-answered questions covering variables such as gender, ethnicity and role of co-morbidities in the outcomes of different ILDs. Furthermore, collaboration with other international registries will enable benchmarking ILD care across countries and continents, enhancing learning in the field of ILD and fostering future research in this important disease area.

Registry	Date	Journal	Publication
UK IPF Registry	2023	European Respiratory Journal	Comparison of antifibrotic availability for patients on the BTS IPF Registry using existing and new prescribing criteria[3]
UK IPF Registry	2021	ERJ Open Research	Idiopathic pulmonary fibrosis in the UK: analysis of the British Thoracic Society electronic registry between 2013 and 2019[4]
UK IPF Registry	2020	European Respiratory Journal	**Abstract:** Eligibility for anti-fibrotic therapy of patients in British Thoracic Society (BTS) UK IPF Registry varies according to the reference values used to calculate FVC[2]
UK Sarcoidosis Registry	2019	BMJ Open Respiratory Research	Sarcoidosis in the UK: insights from British Thoracic Society Registry data[5]
UK IPF Registry	2017	European Respiratory Journal	**Abstract:** First insights from the BTS idiopathic pulmonary fibrosis (IPF) Registry[6]
UK Sarcoidosis Registry	2017	European Respiratory Journal	**Abstract:** Sarcoidosis in the UK: Insights from the BTS interstitial lung disease registry[7]

Table 1: Abstracts and papers published on behalf of the BTS ILD Registry Steering Group since 2013.

1.2 VIEWS FROM REGISTRY STAKEHOLDERS

The BTS Lung Disease Registry Programme is privileged to work with, alongside and in the interests of a variety of stakeholders. The NHS, local clinical teams, hospital managers, trial managers, patients and patient charities are all essential, interconnected parts of the complex tapestry of ILD care. Some of the Registry stakeholders have kindly shared their thoughts about the Registry at this significant milestone:

Participation in the UK ILD Registry provides a way for every respiratory team to demonstrate they are achieving the standards of care set out by NICE for patients with Interstitial Lung Disease. It is driving up national standards of care for our patients.

Dr Lisa Spencer
Consultant Respiratory Physician, Aintree University Hospital
Former BTS ILD Registry Chair and Former BTS Honorary Secretary
Current Member of the NHS Respiratory Clinical Reference Group

In April 2023 Dr Spencer wrote an article for Respiratory Futures discussing the NHSE ILD Quality Dashboard, which is mandatory for English specialist centres[8]. Dr Spencer covered topics including how the dashboard items were chosen, how they can be collated through the UK ILD Registry and how dashboard data may be used to drive improvements in care.

SarcoidosisUK supports the BTS ILD Registry, and we are proud to represent sarcoidosis patients on its Steering Group. The Registry is a vital tool for researchers, medical professionals, and policymakers to better understand sarcoidosis and other ILDs. We urge hospitals and patients to share their data so that the Registry is as comprehensive as possible.

Leo Casimo
Senior Executive, SarcoidosisUK

Action for Pulmonary Fibrosis fully supports the work of the BTS UK ILD Registry and are proud to represent the patient voice on its Steering Committee. For patients, participation in the Registry offers the opportunity, at no additional burden to themselves, to make an important contribution to the development of treatment and care guidelines and to facilitate research to better understand unmet patient needs in ILD. The BTS UK ILD Registry is uniquely positioned to make meaningful inferences, drawing on insights into real world views of variations in clinical practice and patient outcomes to support improvements in NHS service and care pathways. We are encouraged by the Registry's expansion to include all fibrotic-ILDs, which reflects evolving understanding of, and treatment of ILD in recent years. We hope that this will provide richer representation and increased opportunity to improve the lives of all those living with ILD.

Dr Clare Hodkinson
Research Manager, Action for Pulmonary Fibrosis

It is apparent that there are many unexplored correlations between different illnesses which have an influence on the lungs. By collecting as much information as possible on patients, the Registry provides an invaluable and unique resource for researchers. The Registry also keeps track of progression of the disease and could reveal as yet unknown correlations between conditions, treatments and outcomes. The more patients registered, the better will be the understanding of the condition.

Howard Almond
Patient and organiser of support groups

The Registry is more than a tool…it has allowed us to impact patients' lives and improve care provision in the Southwest!

Dr Huzaifa Adamali
Consultant Respiratory Physician, North Bristol NHS Trust

1.3 THE NEW BTS UK ILD REGISTRY (LAUNCHED 2023)

February 2023 was the tenth anniversary of the launch of the UK IPF Registry and the UK Sarcoidosis Registry. This milestone provided the perfect opportunity to discuss the future of lung disease registries at BTS and how they could better serve patients, clinicians and researchers in the future. A number of possibilities were suggested and discussed in detail.

Separately, the National Institute for Health and Care Excellence (NICE) approved the anti-fibrotic nintedanib for use in cases of progressive fibrosing ILD from February 2022 onwards. As this was a new development, it would be important to ensure data are available on the use of nintedanib in this cohort. BTS would be in a unique position to collate data on this important activity nationally, and it would be a missed opportunity if the Registry was not configured to be responsive to novel treatments in ILD.

These developments took place in parallel, and it was decided that the Registry would be both more streamlined and more responsive to changes in the ILD landscape if a new BTS UK ILD Registry was formed. It was agreed that:

- A single 'BTS UK ILD Registry' would be formed, merging the existing databases from the UK IPF and UK Sarcoidosis Registries, as well as expanding to include any other ILD with evidence of fibrosis.

- There would be a 'core dataset' collected for all ILDs, with some additional questions for progressive disease (as well as some questions specific to some diseases). This new dataset would include questions relating to the use of nintedanib.

- The Registry experience would be streamlined for clinicians, with only one interface to navigate. All existing records and user accounts would be retained, and all information relating to NHSE ILD Quality Dashboard returns would remain unchanged. As part of the process the wording of consent forms would also be amended to ensure data linkage could be used to benefit third party researchers going forward.

In short, the new Registry would provide a more streamlined, efficient way to collect and make use of data to benefit a larger group of patients.

Inclusion criteria for the expanded BTS UK ILD Registry

As of 21st February 2023, patients may be included in the new, expanded Registry if they meet all three of the following criteria:

1) First seen in clinic at the participating centre from 1st January 2013.
2) Patients with a new or historic diagnosis of either:
    a. any ILD with evidence of fibrosis, including definite or strongly suspected IPF, *OR*
    b. any sarcoidosis with pulmonary involvement, with or without fibrosis.
3) Informed, written consent has been obtained.

As with the previous UK IPF and UK Sarcoidosis Registries, data may be included both prospectively and retrospectively.

1.4 GET INVOLVED WITH THE BTS UK ILD REGISTRY

The BTS UK ILD Registry is open to recruitment in perpetuity, and there are lots of ways to get involved, whether as a clinician, researcher or patient:

- **I am a clinician…**
  Contact BTS directly at registry@brit-thoracic.org.uk – the team will help you obtain local Caldicott approval and will give you everything you need to get started. If you're based in Scotland, Caldicott approval is already in place nationally through the Scottish Public Benefit and Privacy Panel.

- **I am a researcher…**
  Researchers can apply to access data from the new BTS UK ILD Registry, which includes all records from the previous UK IPF and UK Sarcoidosis Registries. Full details of this BTS Data Access Request Process are available at www.brit-thoracic.org.uk/quality-improvement/bts-clinical-data-policy-and-data-access/.

- **I am a patient…**
  If you're interested in joining the Registry please speak with your consultant, who will be able to let you know if your hospital is signed up. The medical team at your hospital will be able to answer your questions in the first instance, and if you would like anything clarified please do get in touch with the Registry team at BTS.

You can also spread the word about the Registry by sharing Registry reports and other information on social media @BTSrespiratory

PART 2 – The UK Idiopathic Pulmonary Fibrosis Registry

Idiopathic Pulmonary Fibrosis (IPF) is a condition in which progressive scarring of the lungs makes breathing increasingly difficult. It is a devastating disease with a significant symptom burden and poor prognosis. The majority of patients present with breathlessness, cough and fatigue. It has a significant impact on quality of life, affecting physical, emotional, psychological and social aspects of everyday life. For some, scarring of the lungs progresses quickly, while in others it occurs over a longer period of time. We do not yet know why.

Knowledge is power said Sir Francis Bacon, more than 400 years ago. The UK IPF Registry was set up 10 years ago to increase knowledge around the real-world data for people with IPF in the UK. It was envisaged that Registry data would help improve management and service delivery, ensuring the best possible outcomes for people living with pulmonary fibrosis. The Registry has collected health data from 5,052 people affected by IPF across 64 centres, allowing us to look at trends over the last decade.

The COVID pandemic has had significant impact on Registry input, as well as all other aspects of respiratory care. This was reflected in the lower rates of data entry to the Registry in 2020 and 2021. Hopefully this will improve over time as data can be entered retrospectively.

Noticeable trends over time include:

- the increasing age of patients at diagnosis
- a marked drop in lung biopsies for diagnosis
- the largely unchanged time from symptom onset to diagnosis, with 60.0% of patients reporting onset of symptoms more than 12 months, and 36.7% more than 2 years prior to diagnosis
- a significant increase in referrals from secondary care to specialist centres from 50.5% in 2013 to 78.4% in 2023
- diagnosis by MDT occurring in the majority of cases, with only 1 in 33 (3%) cases not discussed in 2022 compared to in 1 in 5.9 (17%) cases in 2013.
- a gradual increase in the proportion of patients deemed ineligible for lung transplantation ‘at any time’ at their first clinic visit, rising from 61% in 2013 to 74% in 2022.
- a consistently low proportion (<10%) of patients recruited to clinical trials at the time of presentation.

Waiting times are an issue throughout the NHS at present but may be particularly noticeable in IPF where most patients are first assessed at a secondary care centre, then wait again for tertiary referral. This is distressing in a condition with such a high symptom burden and poor median survival.

BTS has worked closely with NHSE in producing the NHSE ILD Quality Dashboard items, which use IPF as a surrogate marker for ILD services (as it is the most common ILD). These Dashboard items are designed to mirror the quality statements in the NICE Quality Standard for IPF[1]. The Registry dataset is aligned with the quality statements, facilitating direct benchmarking against these statements. The dashboard was discussed in detail by Dr Lisa Spencer in Respiratory Futures in April 2023[8].

This report shows aggregated national data but can also be used by individual centres to assess their own information. Although dashboard submission is mandatory for English specialist centres, the resources developed to support the collation of dashboard data may be used by any centre – making the Registry directly relevant to assessing services in district general hospitals and in sites across the

devolved nations. The number of participating centres has risen from 25 in 2013 to 87 in 2023, covering 70 Boards/Trusts, which enables the comparison of best practice and patient outcomes across a geographically diverse population.

It is reassuring that the vast majority of patients have their diagnosis made at MDT and are assessed for oxygen, pulmonary rehabilitation and palliative care needs. These metrics have improved over time. Most patients have access to a specialist nurse, although there are still far too many without this service. A minority of patients are offered antifibrotic medications at their first clinic visit. Until this year antifibrotic access in England was limited to patients with IPF who had an FVC of 50-80% predicted. Overall, 37% of patients in the Registry have and FVC above this level at presentation, and this was the major factor in non-prescription of these drugs. Nintedanib has been made available for these patients as of this year and we expect to see a marked change in this in the coming years.

Research is vital to improve our understanding and treatments for IPF. Very few patients are recruited to clinical trials at presentation. This was especially marked during the COVID pandemic, where many trials ceased recruitment. It is particularly exciting that the TIPAL trial (Treating Idiopathic Pulmonary Fibrosis with the Addition of Lansoprazole[9]) is ongoing. This innovative collaboration between the UK IPF Registry and the Norwich Clinical Trials Unit uses the Registry to capture information directly, minimising duplication of data entry. A better understanding of the barriers experienced by patients to participating in clinical trials is required in order to identify the most effective strategies for increasing enthusiasm and opportunities to take part.

As of this year the Registry has now expanded to take on patients not just with IPF but with any other fibrotic lung disease, improving knowledge, data and power for all involved in the care of ILD.

A registry is only as good as the data entered. Most health data held by the Registry are entered by specialist centres. Since the Registry was launched 69% of patients were referred from secondary care respiratory teams. We would encourage other centres to become involved in the Registry – anyone who is interested should contact registry@brit-thoracic.org.uk

Inclusion criteria for the UK IPF Registry

These were the inclusion criteria for the UK IPF Registry until February 2023. Participating centres were asked to enter data on patients who met 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.

New UK ILD Registry 2023

The UK ILD Registry expanded in February 2023 and now includes all fibrosing ILD and non-fibrosing sarcoidosis.

The current, updated inclusion criteria are listed on page 12 along with details of how to take part.

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 from January 2013 to 19th February 2023

At 19th February 2023, 81 centres had approval to participate in the UK IPF Registry, with 64 centres having contributed clinical data. The full list of 87 centres which currently have approval to participate in the Registry is given on page 35.

5,052 patient demographic records

4,790 complete clinical/diagnosis records from the first clinic visit

3,523 follow-up records representing 1,516 unique patients. These follow-up records represent 30% (1,516/5,052) of all records, with patients where follow-up data have been entered. Patients have a mean of two follow-up records each.

2.1 THE IPF PATIENT COHORT

Demographic information is collected at the first clinic visit.

Sex

The proportion of male and female patients has remained consistent over time, with a mean of 22.2% (1,084/4,886) females and 77.8% (3,802/4,886) males over the lifetime of the Registry. These data are in keeping with the known epidemiology of this disease.

Age

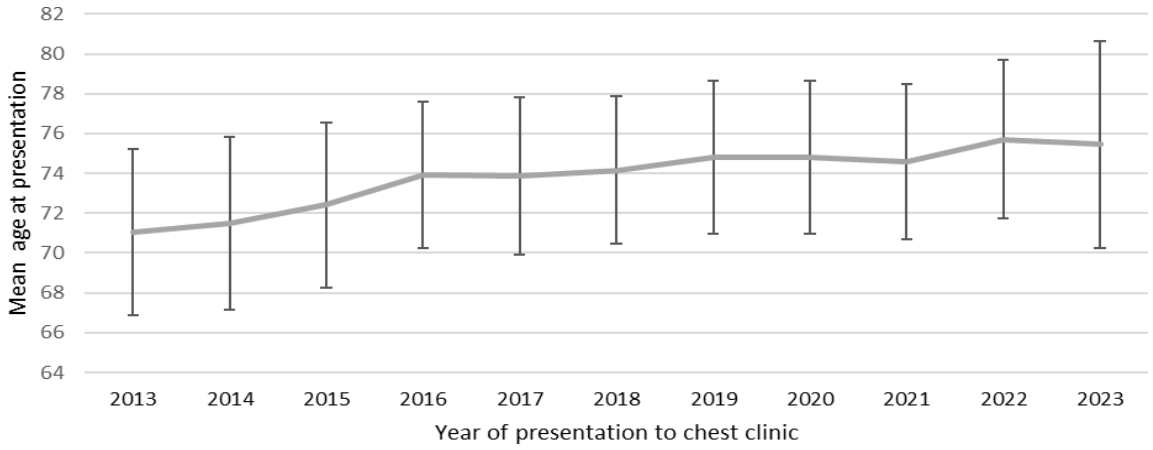


Figure 1: Mean age at presentation over time

The mean age of patients at presentation (shown here with error bars representing standard deviation) gradually increased over time from 71.0 years in 2013 to 75.7 years in 2022. Over the lifetime of the Registry the mean age was 74.0 (± *8.1*), with 73% (3,524/4,82) aged 70 or over.

Comorbidities

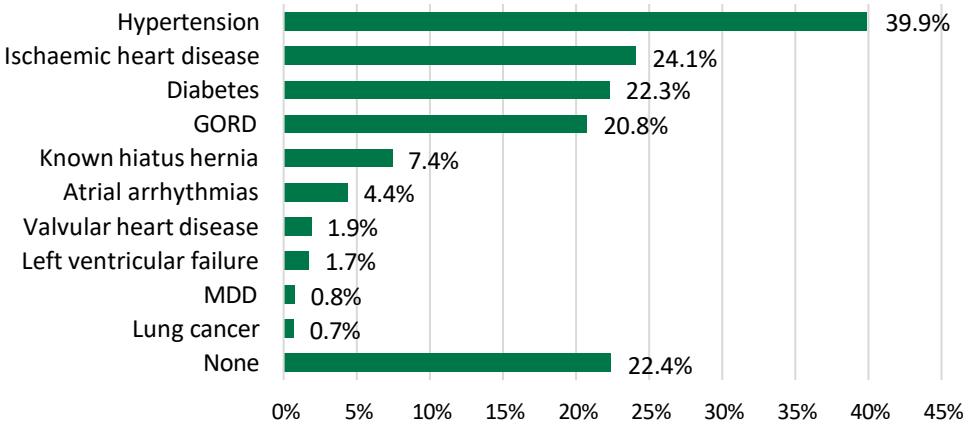


Figure 2: Comorbidities at presentation

Patients with IPF have a high burden of comorbidities at presentation, with 76% having at least one comorbidity. The nature and proportion of comorbidities remain stable over the lifetime of the Registry.

Smoking

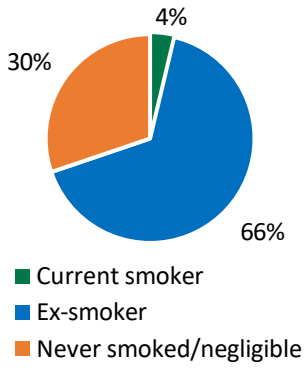


Figure 3: Smoking status at presentation
The proportion of patients who were smokers at presentation has remained consistent through the lifetime of the Registry. Overall, 4% (159/4,281) of patients were current smokers, 66% (2,829/4,281) ex-smokers and 30% (1,293/4,281) had never smoked or smoked a negligible amount (<5 pack years).

Relatives with IPF

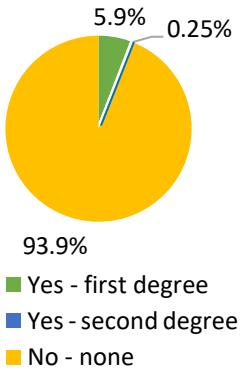


Figure 4: Known relatives with IPF
Where clinicians had recorded family history, approximately 5.9% (188/3,213) of patients had a first degree relative known to have IPF and 93.9% (3,017/3,213) had no known relatives with IPF. Very few patients reported a second degree relative with IPF. These figures 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

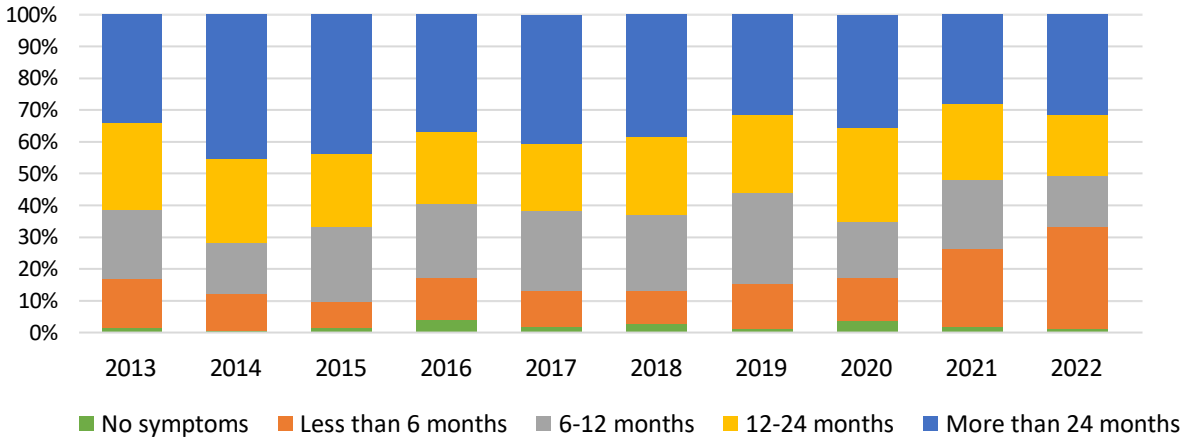


Figure 5: Duration of IPF symptoms prior to presentation over time
Unfortunately, patients often have symptoms for a considerable period prior to diagnosis. Since 2013, 60% (2,561/4,265) of patients reported having chest symptoms for more than 12 months before their first hospital clinic visit.

Route of referral

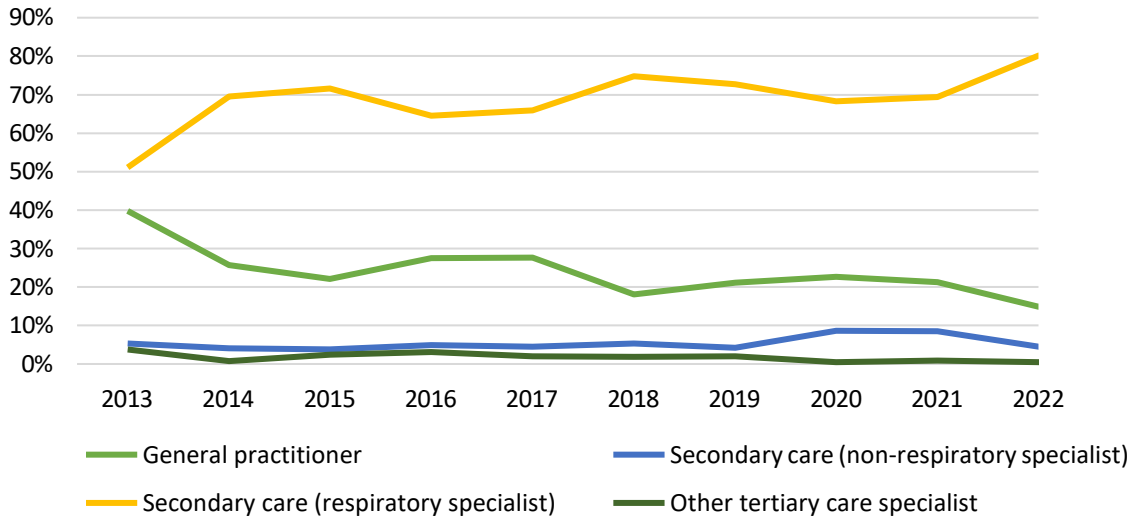
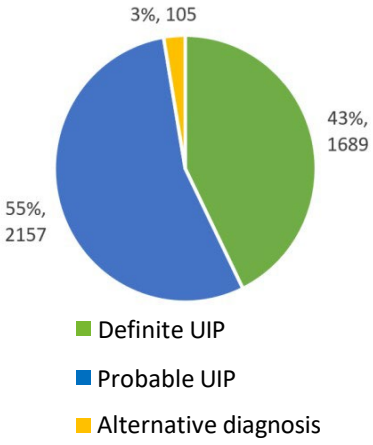


Figure 6: Route of referral to clinic over time
Throughout the lifetime of the UK IPF Registry the primary route of referral has been directly from a respiratory specialist in secondary care. Since 2013 there appears to have been a significant increase in referral from respiratory specialists in secondary care (from 50.5% in 2013 to 78.4% in 2022 – only 2 months of data are available for 2023) and a comparable reduction in referral from general practice (from 39.4% in 2013 to 14.5% in 2022). This may be due to the introduction of antifibrotic therapy – for which referral to tertiary care is required – and may have an impact on waiting times.

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



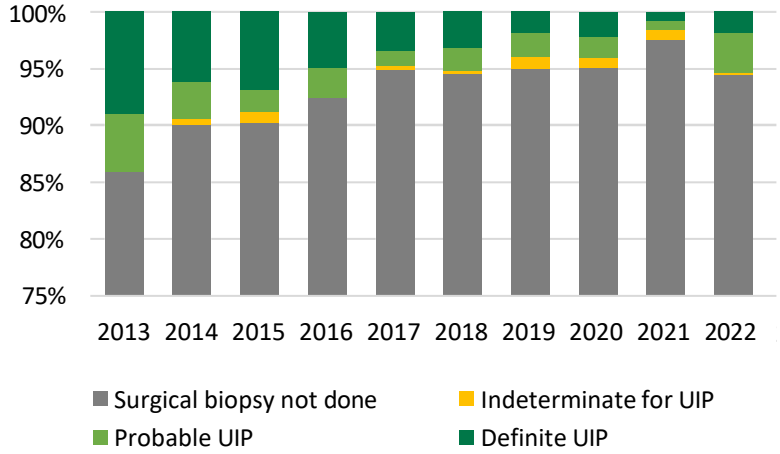


Figure 7 (left): **HRCT pattern at presentation, and**
Figure 8 (right): **Surgical biopsy data at presentation over time**
The majority of patients are diagnosed using high-resolution computed tomography (HRCT) scanning. Roughly equal proportions of these scans reported patterns in keeping with definite 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 (8%, 282/3,928). The percentage of cases where a biopsy was conducted decreased from 14% in 2013 to 5.5% in 2022, especially since the diagnostic guidelines were updated in 2018.

Patient Waiting Times

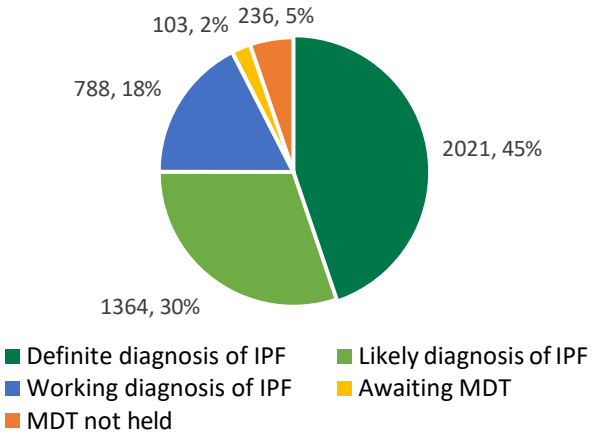
Mean waiting times from referral to first clinic visit were 13.6 weeks (13.9 for English specialist centres and 12.8 for other centres). Mean waiting times from referral to multidisciplinary team (MDT) meetings were 12.8 weeks (12.5 and 13.8 for English specialist centres and other centres respectively).

The low proportion of referrals from general practice may negatively impact wait times in a way which is not reflected here. Wait times following referral from secondary care do not take into account the initial wait time for referral from general practice to secondary care.

MDT

Figure 9: Outcome of MDT
The majority of cases are discussed at MDT, with 93% having been discussed at the first clinic visit and a further 2% 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 17.0% (28/165) of cases in 2013 to 3% of cases in 2020 (19/619).



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

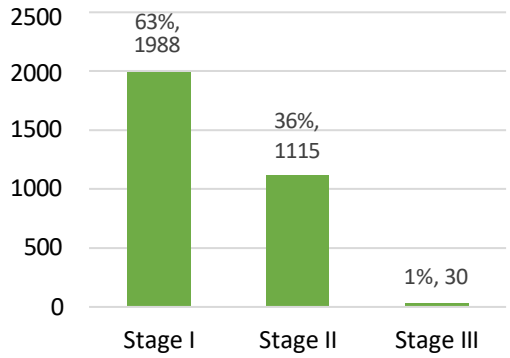


Figure 10: GAP stage at presentation
At first presentation the overwhelming majority of patients with IPF are categorised as having GAP Stage I or Stage II disease (99% of patients overall – 3,103/3,133). GAP staging is a marker of IPF disease severity, calculated using sex, age and lung function details[10].

Pulmonary hypertension

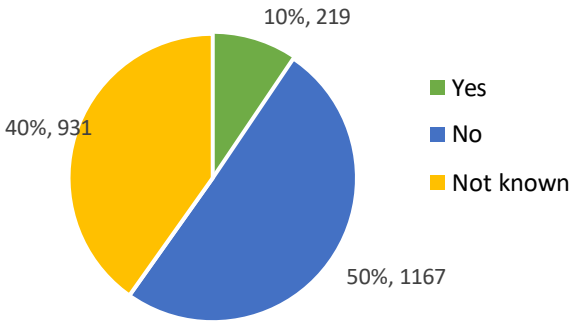
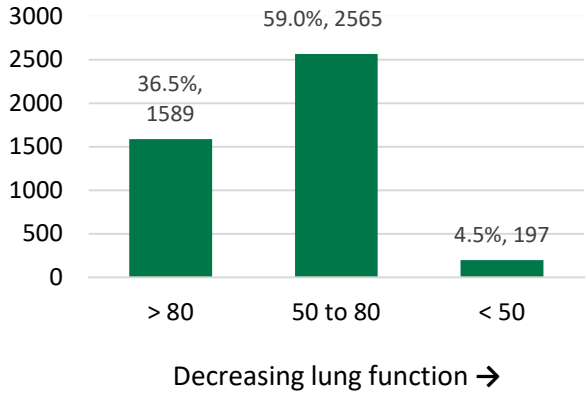


Figure 11: History of pulmonary hypertension
Overall 10% (219/2,317) of patients at presentation had previously been confirmed to have pulmonary hypertension or right heart strain, secondary to their lung disease, confirmed on echocardiogram or right heart catheterisation.

Lung Function (at presentation)



NICE Guidance Changed in 2023

In February 2023 NICE removed the requirement for patients to have an FVC below 80% predicted before nintedanib could be prescribed.

For full details see NICE Technology Appraisal Guidance TA864[11].

Figure 12: Forced vital capacity (FVC) at presentation, and
At entry 37% of patients have an FVC over 80% predicted and therefore were above England's National Institute for Health and Care Excellence (NICE) criteria for antifibrotic treatment for a number of years.

Until recently in England, Wales and Northern Ireland patients with FVC values <50% predicted were less likely to be referred on to a specialist centre, as treatment could not 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. It may be that this pattern of reported lung functions shifts over the coming years in direct response to the changed guidance.

Drug treatment at presentation

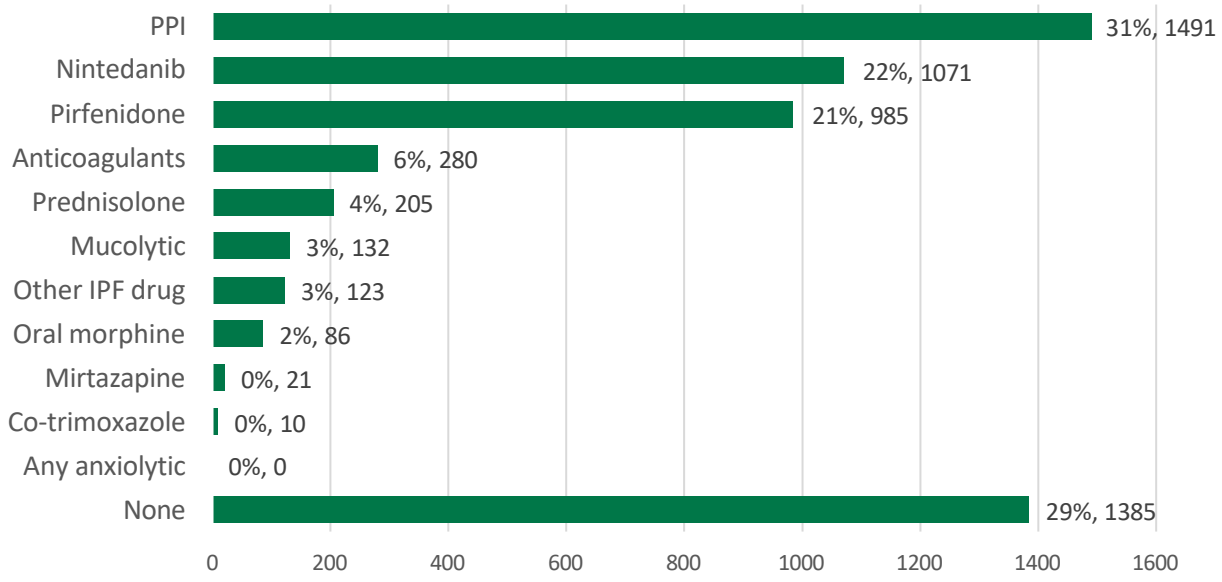


Figure 13: Drug treatment at presentation (including treatment prescribed at first clinic visit)
These data need to be interpreted carefully as they reflect drug use over the lifetime of the UK IPF Registry. Pirfenidone has been available through the NHS since 2013, whereas nintedanib has only been available 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

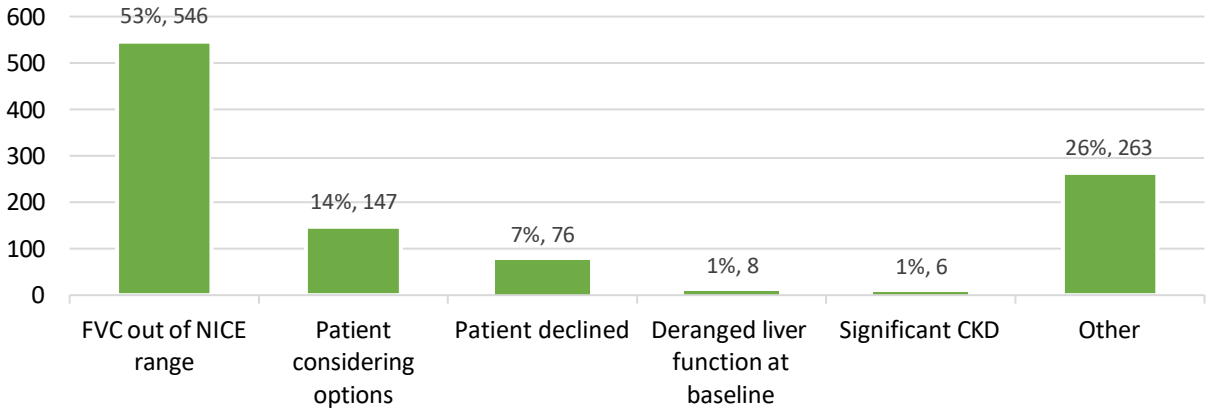


Figure 14: Reason for not starting antifibrotic treatment at first clinic visit

Since 2013, 49% of patients were not prescribed antifibrotic treatments at presentation. Until 2023 suitability for drug treatment was partly based 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 over half of patients (53%) this was because the FVC was outside the range then approved by NICE. For only 14% 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.

Oxygen Therapy

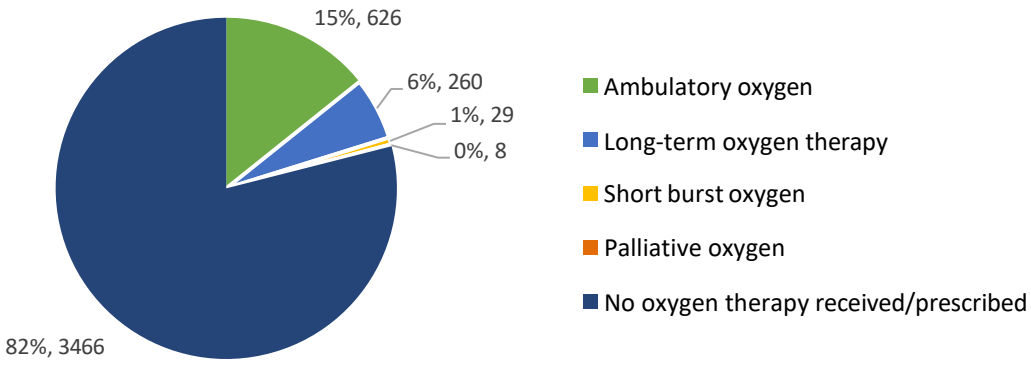


Figure 15: Oxygen therapy at presentation

At presentation 90% (1,744/1,937) of patients had their oxygen needs assessed, with the majority (60%, 1,0471,744) not requiring oxygen therapy at that time. One in eight (16%, 647/4,113) patients were receiving or newly prescribed at least one form of oxygen therapy at their first clinic visit.

Inclusion in Clinical Trials

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; where known, 7.5% of patients (272/3,603) were recruited to a clinical trial at presentation.

Lung Transplantation

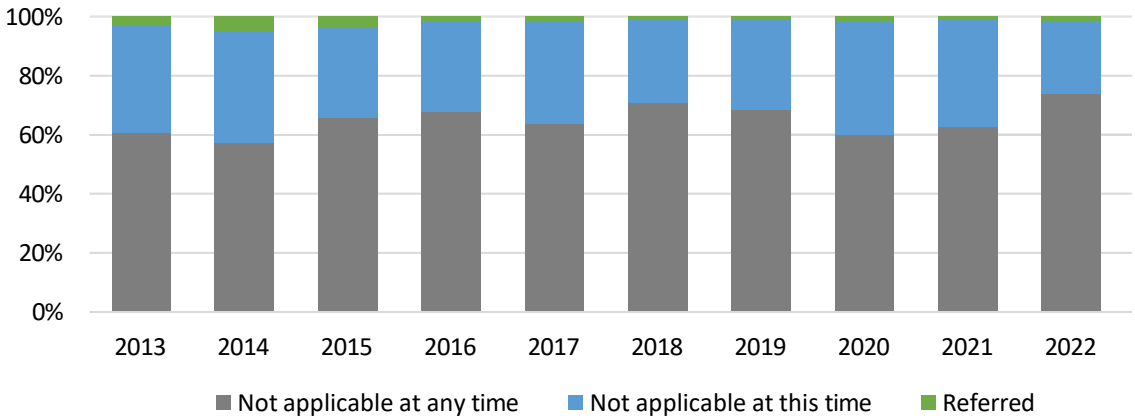


Figure 16: Referral for lung transplantation at presentation over time

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 may have increased gradually, rising from 61% in 2013 to 74% in 2022 (data for 2023 represent only the first two 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 1, page 16).

Patient Mortality

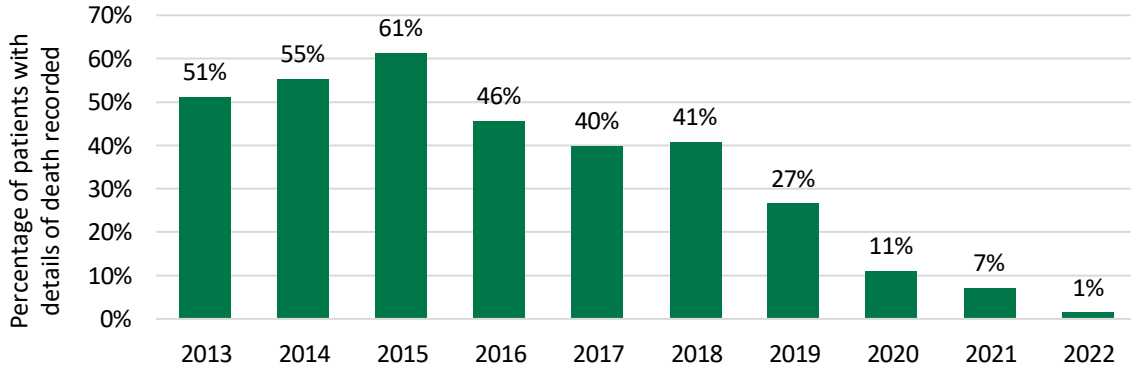


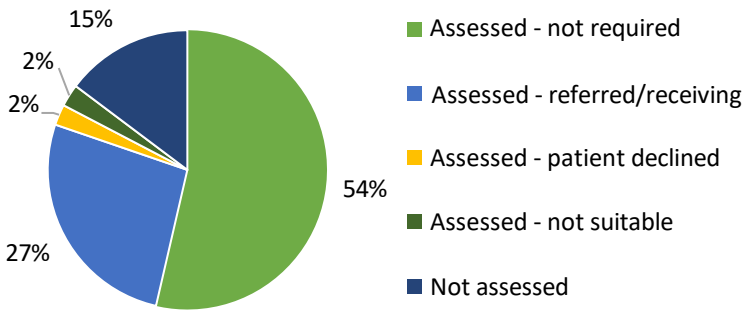
Figure 17: Patient mortality by year of presentation

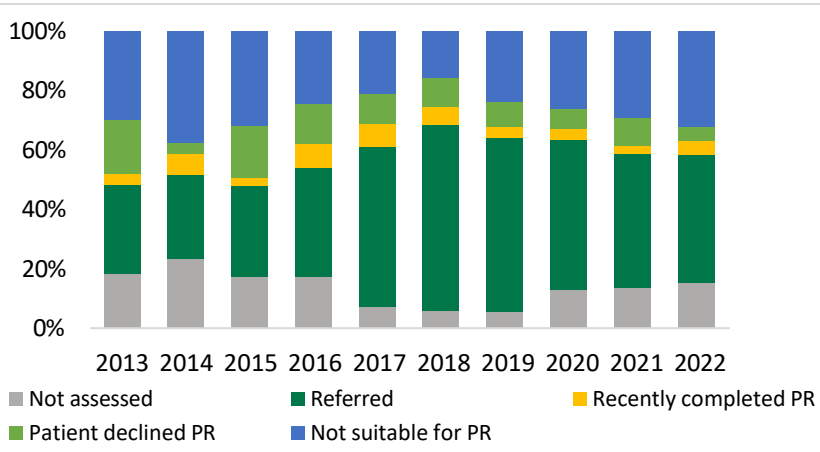
The overall IPF mortality rate for the lifetime of the Registry is 31%. This chart breaks that figure down, showing the mortality rates to date of patients who first presented to the treating centre each year. These data should be interpreted with caution for a number of reasons, including:

- The patients included in the Registry are overwhelmingly (approximately 90%) entered from specialist, prescribing centres in England. This means the population may be skewed toward a different patient group to those seen exclusively on secondary care.
- 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: **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 93% of cases have already been discussed at MDT by the time of the first clinic visit and a further 2% due to be discussed at an upcoming MDT. See Figure 9, page 19.
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 89% (1,967/2,211) 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 18: 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, 85% (1,347/1,580) of follow-up visits included an oxygen needs assessment. Of those assessed, the majority did not require oxygen therapy at the time (63%, 847/1,347). This question was added to the UK IPF Registry dataset in December 2019, therefore the number of responses is comparatively low.

Quality Statement 4: **Pulmonary rehabilitation (PR) programmes provide services that are designed specifically for IPF.**	 **Figure 19: 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, 89% (2,494/2,792) of patients had their PR needs assessed and 11% (298/2,792) did not. Of those assessed the majority (57%, 1,426/2,494) were referred for PR. There was a large increase in the proportion of patients referred for PR over time, from 15% of all patients in 2013 to 54% in 2018. This then decreased to 35% by 2022, possibly as a result of 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, 28% (687/2,494) 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 82% (2,444/2,993) 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 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 71% in 2013 to 88% in 2019. However, as above there was greater variability from 2020-22 (82%, 88% and 80% respectively), possibly due to changes in practice resulting from the COVID-19 pandemic.

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.

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 10% 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[12] and these individuals may be underrepresented in the database.

Data collected from 50 centres over the lifetime of the Registry (January 2013 to 19th February 2023) indicate that sarcoidosis predominantly affects women and presents most commonly between the ages of 41 to 60 years (53% of patients). The majority of people were never smokers with only 7% 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 38% of cases in 2013 rising to 64% in 2022.
- 10% of patients are receiving immunosuppressive therapies (other than prednisolone).
- Only 2% of patients are recruited into clinical trials.
- Few patients (24%) are provided information about other services, such as patient support groups, helplines or mental health services.

Inclusion criteria for the UK Sarcoidosis Registry

These were the inclusion criteria for the UK Sarcoidosis Registry until February 2023. Participating centres were requested to enter data on patients who met 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.

New UK ILD Registry 2023

The UK ILD Registry expanded in February 2023 and now includes all fibrosing ILD and non-fibrosing sarcoidosis.

The current, updated inclusion criteria are listed on page 12 along with details of how to take part.

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 from January 2013 to 19th February 2023

At 19th February 2023, 81 centres had approval to participate in the UK Sarcoidosis Registry, with 50 centres having contributed clinical data. The full list of 87 centres which have approval to participate in the Registry is given on page 35.

937 patient demographic records

839 complete clinical/diagnosis records from the first clinic visit

446 follow-up records representing 218 unique patients.

3.1 THE SARCOIDOSIS PATIENT COHORT

Patient demographic information is collected at the first clinic visit.

Gender

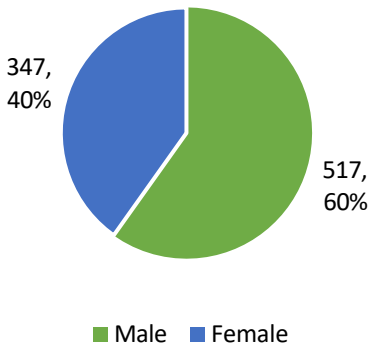


Figure 20: Sex of patients
The proportion of male (60%, 517/864) and female (40%, 347/864) patients has remained consistent over the lifetime of the UK Sarcoidosis Registry.

Age

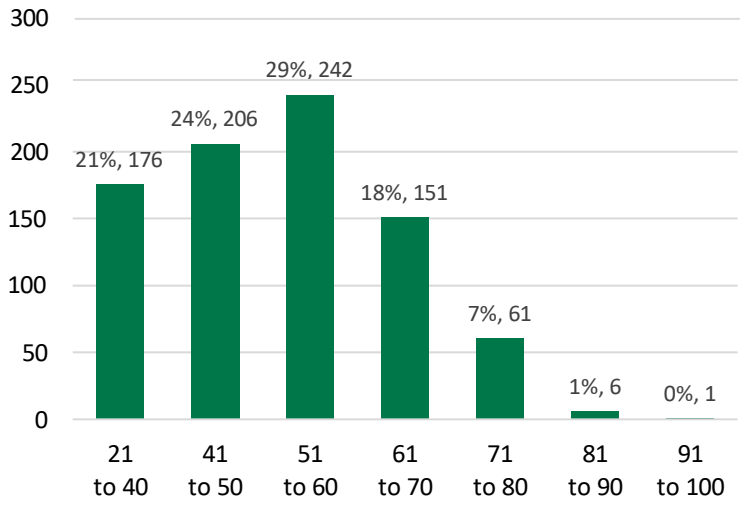


Figure 21: Age at presentation
Just over a quarter of patients (26%, 219/843) presented over the age of 60, with the mean age at presentation being 52.7 years, with a standard deviation of ± 26.9.

Ethnicity

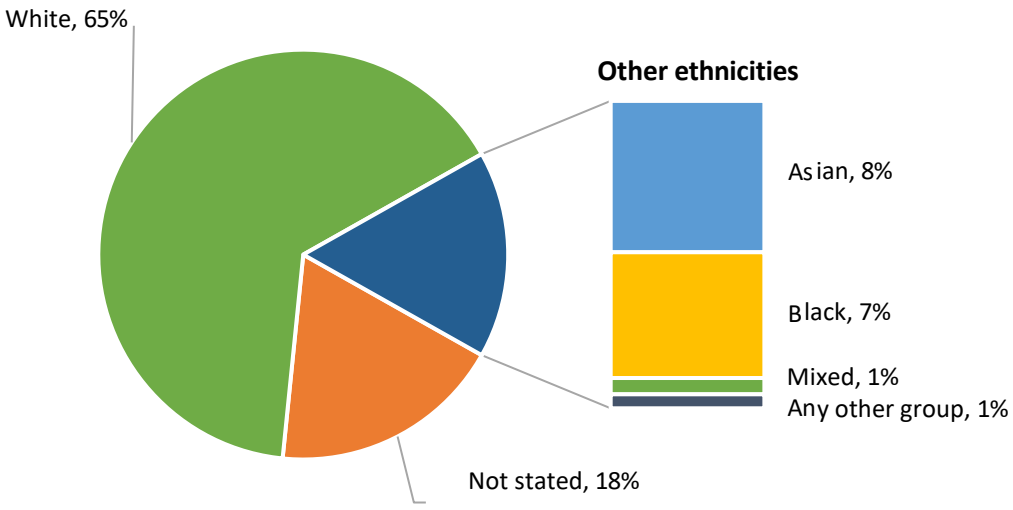


Figure 22: Patients by ethnic group
The majority of patients in the Registry were white (65%, 527/808). Although sarcoidosis in known to be more prevalent in black populations, only 7% (54/808) of patients were black. This figure likely reflects the populations from which Registry data were obtained, 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[13].

Smoking status

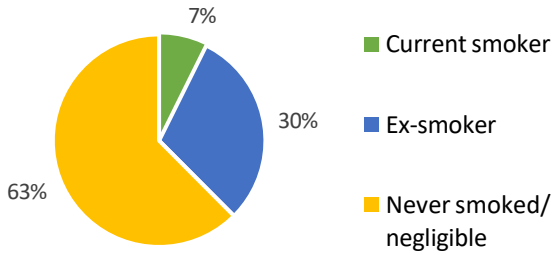


Figure 23: Smoking status at presentation
Overall 37% (256/682) of patients were either smokers or ex-smokers at presentation (patients were counted as ex-smokers if they quit more than three months before their first clinic visit).

62% (426/682) 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 (64%, 383/598) had no reported comorbidity at current presentation.

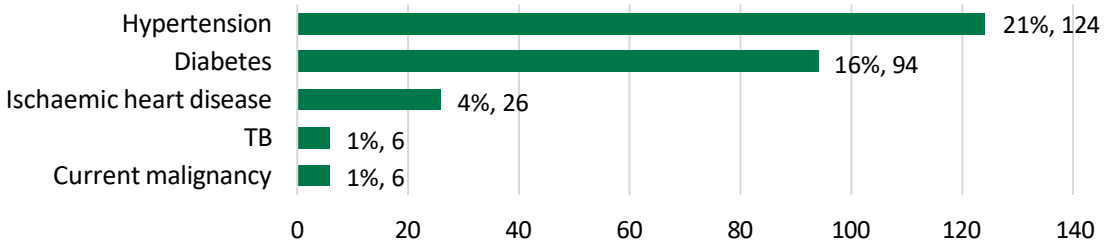


Figure 24: Patient comorbidities at presentation
The most commonly reported comorbidities were systemic hypertension (one in five patients – 21%, 124/598) and diabetes (16%, 94/598). These conditions are highly prevalent in the general population. Where at least one comorbidity was recorded patients had a mean of 1.2 reported comorbidities each.

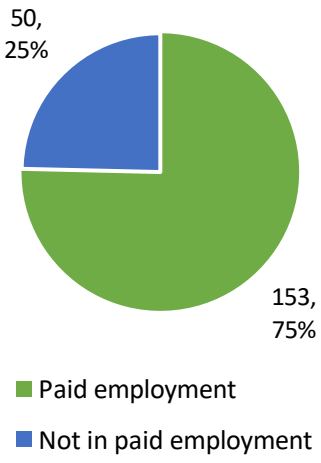
Relatives with sarcoidosis

A minority of patients in the UK Sarcoidosis Registry are known to have relatives who have been diagnosed with sarcoidosis. Overall, 4% (17/478) 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 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 25: Employment status at presentation
Where employment status at presentation was known, three quarters (75%, 153/203) 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

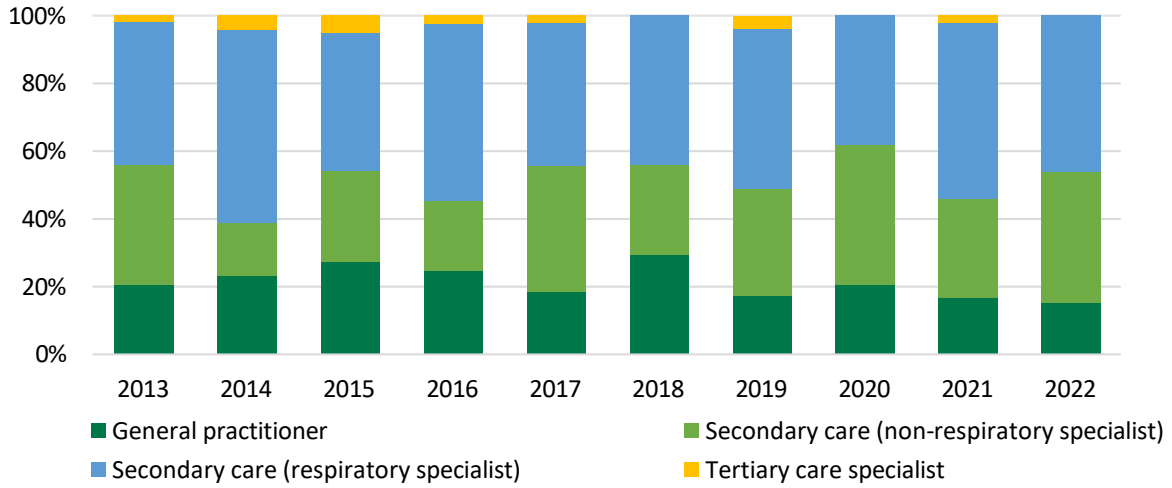


Figure 26: Route of referral to clinic over time

Overall, 46% (364/792) of patients were referred from respiratory physicians in secondary care. Referrals from general practice have remained low at 23% overall (183/792), 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% (269/725) of cases known to have been diagnosed incidentally.

Symptoms at first clinic visit

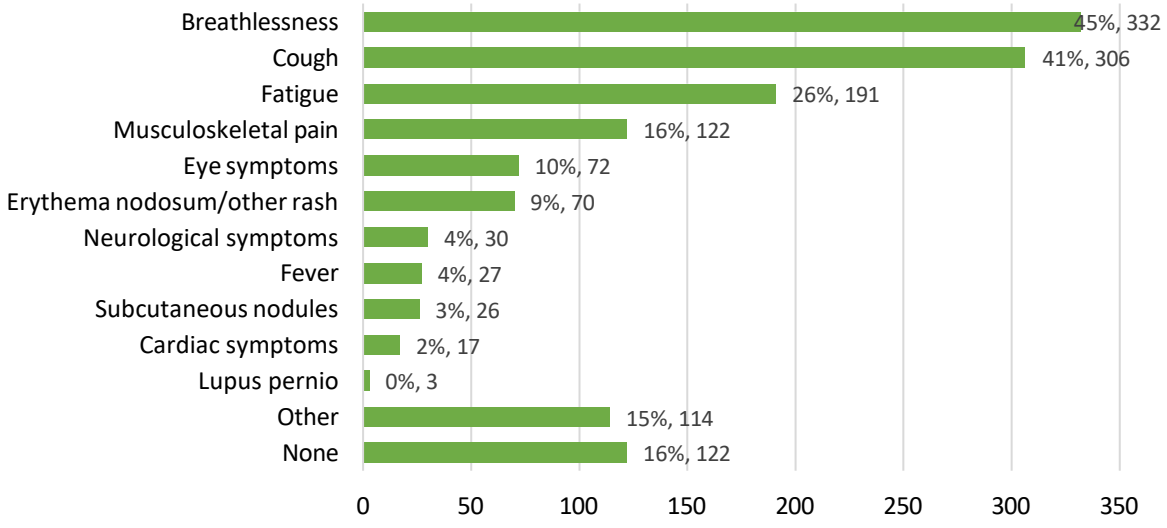


Figure 27: Symptoms reported at first clinic visit

The most common symptoms were breathlessness (45%), cough (41%) and fatigue (26%). Musculoskeletal pain (16%), eye symptoms (10%) and skin rashes (9%) 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 93% (564/609) 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.

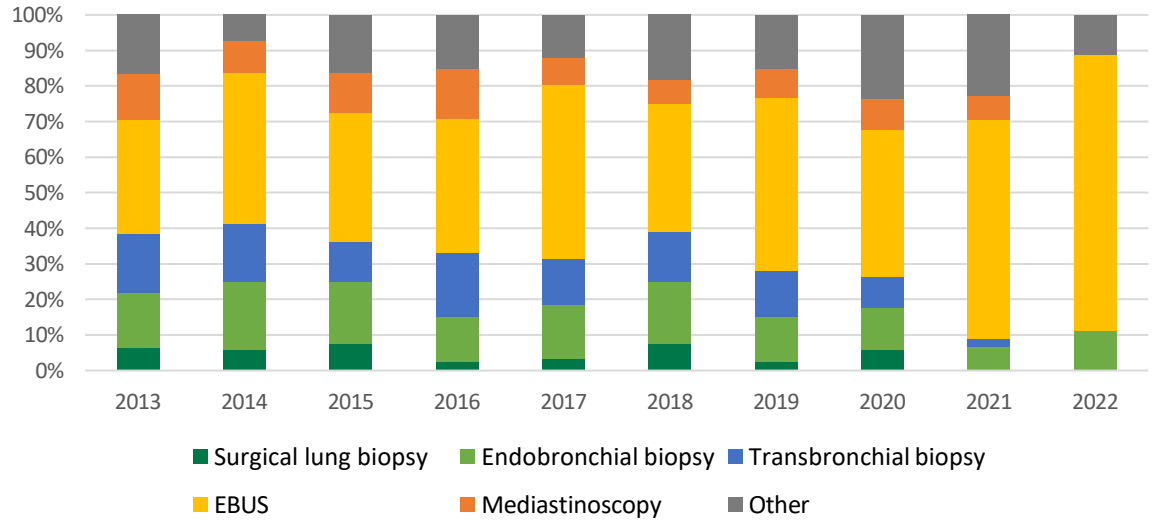


Figure 28: Biopsy techniques used at presentation over time
A number of different biopsy techniques were employed. Endobronchial ultrasound (EBUS) was consistently the most popular, involved in 52% (293/564) of cases where at least one biopsy was conducted. In the first two months of 2023 only three responses were received for this question.

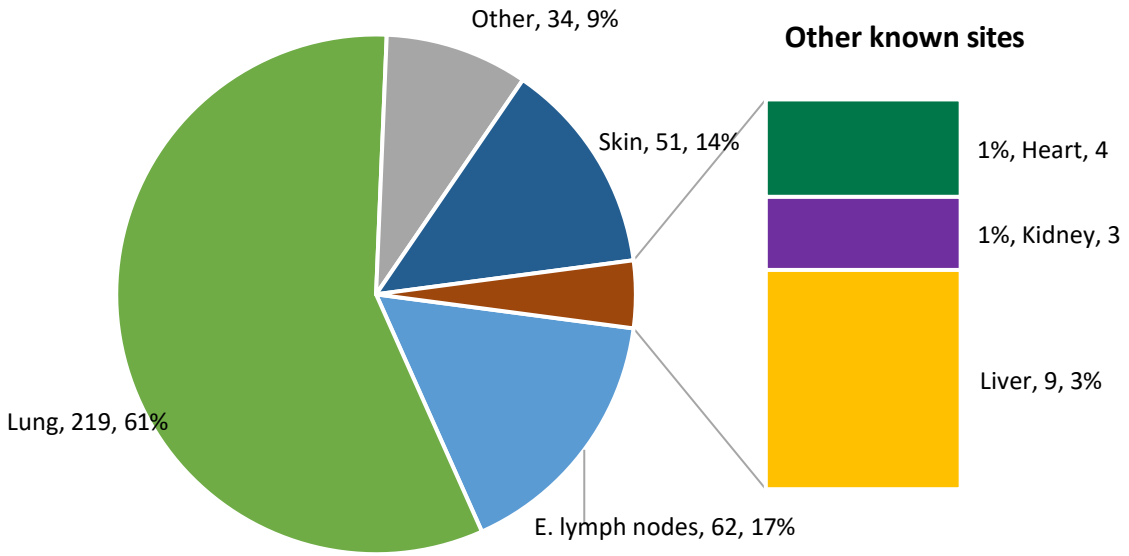


Figure 29: Site(s) from which histology obtained at presentation
The most common biopsy sites were lung (61%, 219/359), extra-thoracic lymph nodes (17%, 62/359) and skin (14%, 51/359). The variety of biopsy sites investigated is indicative of the multisystem involvement commonly observed in sarcoidosis.

3.3 CLINICAL DATA AT PRESENTATION

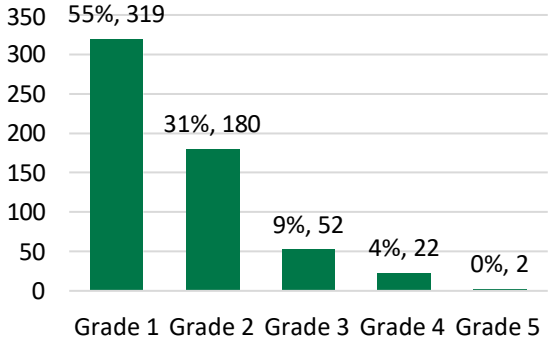
The data presented in this section are collected once for each patient, at the first clinic visit.

Pulmonary hypertension

At presentation 1% (4/269) 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.

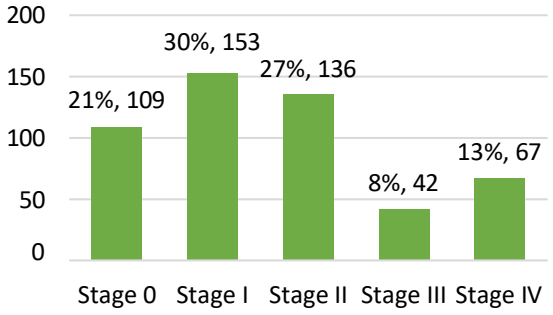
MRC dyspnoea scale

Figure 30: Breathlessness at presentation
Most patients (87%, 499/575) 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 31: Chest radiograph at presentation
At presentation the majority of patients were observed to have either Stage I with lymphadenopathy only (30%, 153/507) or Stage II lymphadenopathy and parenchymal involvement (27%, 136/507) disease according to the Siltzbach sarcoidosis classification system[14].



Blood tests

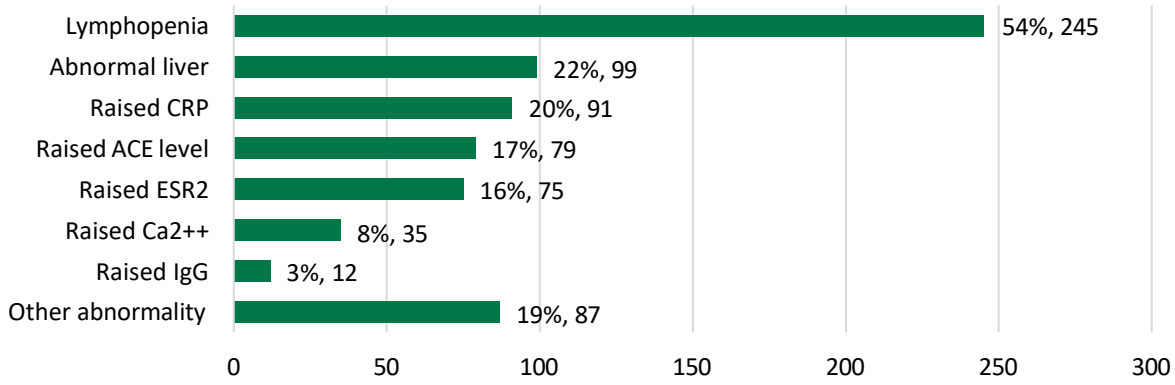


Figure 32: Blood test abnormalities recorded at presentation
The most common abnormality recorded in blood tests at presentation was lymphopenia, identified in 54% (245/456) of patients. Raised angiotensin converting enzyme (ACE) levels and calcium levels (as defined at local centres) were reported in 17% and 8% of cases respectively.

HRCT pattern

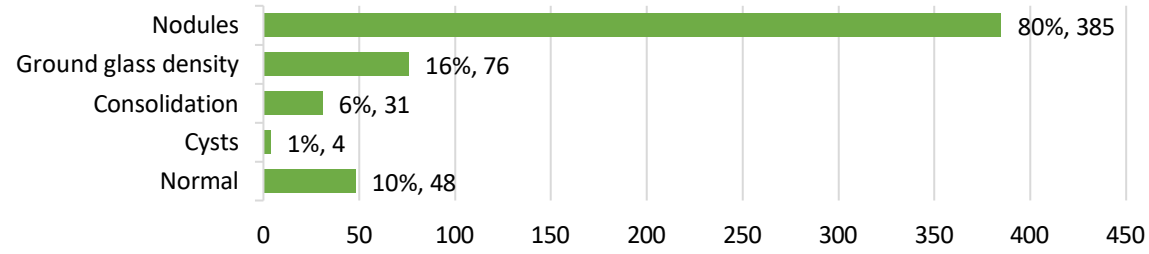


Figure 33: Parenchymal abnormalities identified on HRCT at presentation
The most common parenchymal abnormality identified on HRCT imaging at presentation was nodules, found in 80% (385/480) of cases. HRCT imaging was found to be normal in 10% (48/480) of cases.

Current drug treatment

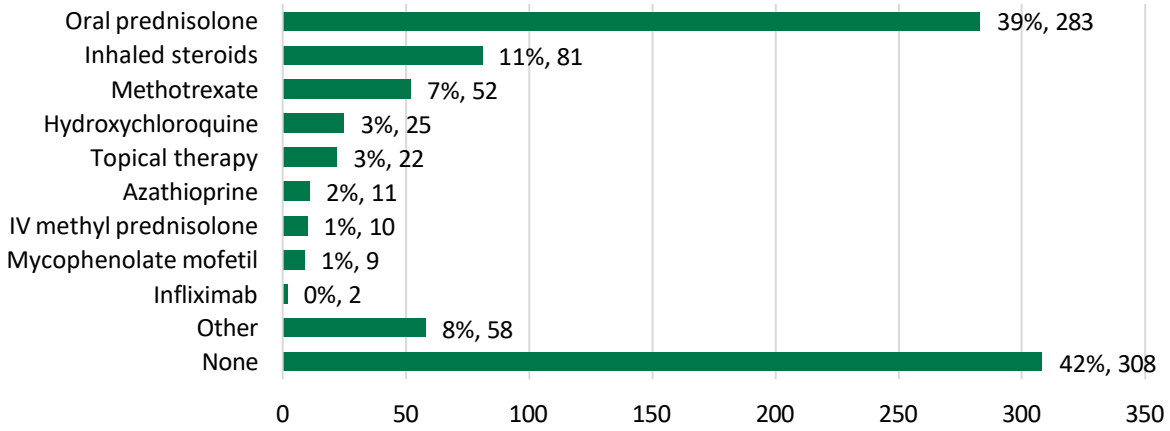


Figure 34: Drug treatment at presentation
The majority of patients were either not started on treatment (42%, 308/730) or managed with systemic corticosteroids (46%, 339/730). A number of alternative agents were used, with none used in more than 11% of cases. This broadly reflects previous BTS guidance on the management of sarcoidosis[15].

Referral to other services

Only 24% (64/264) of patients were referred or signposted to other services at the time of presentation. By other services we mean services providing either other clinical support or non-clinical support. Of those who were, 50% (32/64) were given details for patient support groups and 14% (9/64) for helplines, and 6% (4/64) were referred to or informed of mental health support.

Inclusion in clinical trials

At presentation only 1.9% (13/678) of patients were recruited to a clinical trial. There is a need for more clinical research in sarcoidosis.

PART 4 – Ten Year Forward Vision for the UK ILD Registry

The ten-year report demonstrates the impact of this nationwide registry in monitoring presentations and outcomes in idiopathic pulmonary fibrosis (IPF) and sarcoidosis. The Registry offers value to clinical services in maintaining the highest levels of care and the potential to translate the data into tangible benefits to patients. In order to achieve such benefits, thereby maximising the contributions from services and patient-level data, we propose a ten year forward vision which includes greater completeness, effective integrations, and development of robust outputs.

Moving forward, our vision is that the data collected within the Registry is more generalisable and more complete; at present, the majority of the data arises from a core of specialist centres. Although we envisage that specialist centres will be the main contributors to the Registry, we look forward to the continued increasing involvement of non-specialist centres. We expect that this will increase the number of patients with follow-up data and the number of follow-up data-points per patient, with the resultant ability to have greater understanding of trends and changes over time. We hope that the initiatives to support data entry will catalyse a greater representation of patients recorded in the Registry, such as the such as the data input personnel funded by Boehringer Ingelheim in some centres.

The vision to make the Registry relevant to all people with Interstitial Lung Disease (ILD) is ongoing and we anticipate it will continue. ILD encompasses a large number of diagnoses, and this report demonstrates a successful merge of the UK ILD and UK Sarcoidosis Registries to support awareness of similarities in presentation and outcomes. Heterogeneity in ILD aetiology, and changes in management guidance according to progressive lung fibrosis, highlight an opportunity for the forward vision to interpret specific disease incidence over time and to report the ways in which clinical data collected in the Registry are different or similar according to ILD diagnosis.

To support reliable interpretations of care and outcomes, as well as provide a rich dataset for secondary use, completeness of data should be a key aim in the ten-year vision. The Registry requires a minimal core dataset and tolerates incomplete data collection per patient record, but we envisage as the Registry expands data completeness will increase. Barriers to record entry should be identified, and supporting methods of facilitating data entry is a key aim of the Registry. This aim is ongoing, including the revision of the data input and collection system, supporting standardisation and the minimisation of resource requirements. A more complete dataset will provide greater ability to explore the benefits and harms of disease modifying therapy, as well as understanding the reasons for uptake of transplant and palliative care interventions. Greater integration of the Registry within clinical care will provide more benefit to patients and more complete datasets. Our vision is that in the future there will be more automated data capture processes from electronic health record data and that capture of data will be better integrated into routine care.

The ability to undertake record linkage should maximise the potential of benefit from the Registry. Linkage to external datasets is possible for records where appropriate consent is in place, and as the numbers increase so does opportunity for insight. Linkage to, or integration with, future databases containing patient reported outcomes, relative and carer data, and/or socioeconomic data will contribute to the understanding of disease burden nationally. Digital health technologies hold exciting promise in chronic progressive lung disease to support patient accessibility of services and engagement with their own healthcare. This can include registration within digital health portals to facilitate patient-clinician interactions, as well as the use of remote devices to support disease monitoring. The forward

vision of the Registry can help report how integration of such technologies may impact upon healthcare over the next ten years by capturing whether patients and services engage in usage.

Accessibility to, and participation in, clinical trials is a vital opportunity for all individuals with ILD to potentially benefit from novel therapies and improve disease outcomes over time. At presentation, the numbers of individuals being enrolled to clinical trials has been consistently low, at least in part due to the incomplete follow-up data collection. These individuals may go on to enrolment at follow-up visits, or may not meet trial inclusion criteria. The Registry should continue to support the delivery of research within the UK. The TIPAL study[9] utilises the Registry to capture trial data required, as a way of maximising resource and avoiding duplication, and we envisage further similar initiatives.

As data included in the Registry continue to grow, the Registry will prove to be a vital resource for clinical services, including the development of dashboards to monitor management and representative populations for clinical researchers to address key priorities through data access requests. Optimising data capture and data management is a necessary process to ensure research integrity and data quality is maintained. Our vision is that greater integration, collaboration and outputs on a personal, centre and national level will lead to improved patient care.

PARTICIPATING SITES

The following organisations participate 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
Charing Cross Hospital, Imperial College 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
Doncaster Royal Infirmary, Doncaster and Bassetlaw Teaching Hospital NHS 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
Kingston Hospital, Kingston Hospital NHS Foundation Trust
Lister Hospital, East and North Hertfordshire NHS Trust
Liverpool Heart and Chest Hospital, Liverpool Heart and Chest Hospital NHS Foundation Trust
Medway Maritime Hospital, Medway 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, Guy's and St Thomas' 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 Bartholomew's Hospital, Barts Health NHS Trust
St James' University Hospital, The Leeds Teaching Hospitals NHS Trust
St Mary's Hospital, Imperial College Healthcare NHS Trust
Torbay Hospital, Torbay and South Devon 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
Whiston Hospital, St Helens and Knowsley Teaching Hospitals NHS 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
Monklands District General Hospital, NHS Lanarkshire
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
Morriston Hospital, Swansea Bay University Health Board
University Hospital Llandough, Cardiff and Vale University Health Board
Wrexham Maelor Hospital, Betsi Cadwaladr University Health Board

Northern Ireland

Altnagelvin Area Hospital, Western Health and Social Care Trust
Antrim Area Hospital, Northern Health and Social Care Trust
South West Acute Hospital, Western 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 UK 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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