BTS ILD Registry Annual Report 2019

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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

This is the fourth annual report from the BTS Interstitial Lung Disease Registry, which includes the UK IPF Registry and the UK Sarcoidosis Registry.

As it enters its sixth year the BTS ILD Registry is going from strength to strength. The growing number of patient records and of sites contributing to the Registry are a testament to the commitment and quest for accurate knowledge and insight from the UK ILD community. At 31st October 2019 the BTS ILD Registry contains over 2,470 complete IPF cases (with longitudinal data for over 700 unique patients) and over 490 sarcoidosis cases (with longitudinal data for over 80 patients). According to published data this makes the UK IPF Registry one of the largest and most comprehensive longitudinal IPF registries in the world from a single country[1].

Contribution to the BTS ILD Registry has been voluntary to date. Pressures in the NHS remain high, with insufficient resources for most sites to capture the majority of cases on a voluntary basis. Disease registries with high capture rates are known to play crucial roles in healthcare, as demonstrated by those for pulmonary hypertension and cystic fibrosis. Such registries facilitate clinical trials design, patient identification for recruitment, research, and measuring and improving clinical outcomes for patients.

With this in mind NHS England (NHSE) will make completion of the UK IPF Registry mandatory for England's Specially Commissioned Regional ILD Centres. UK IPF Registry participation will be written into the ILD service specification that is contractually agreed with NHS hospitals providing these services, and it is estimated that this action will permit capture of >60% of all new IPF cases in England going forward. This will undoubtedly generate a powerful database and open up avenues of research previously out of reach using small datasets from individual sites. Data submission to the UK Sarcoidosis Registry, as well as submission to the UK IPF Registry from district general hospitals, is still very much encouraged although these submissions will remain voluntary.

Sites maintain full ownership of data from their own hospital and can collaborate with other ILD centres on projects following relevant local ethical approval. In addition, in January 2020 BTS will open its data access request process, allowing organisations to apply to access data from BTS programmes – including the UK IPF Registry and the UK Sarcoidosis Registry – for the purpose of research. This undoubtedly will open up new possibilities for UK researchers. Data submission to the BTS ILD Registry also provides hospitals with an easily searchable dataset to identify potential patients who may be suitable for clinical trials. Sites can access their cases at any time and search for specific parameters which may form part of clinical trial inclusion criteria, e.g. spirometry values.

Discussions between BTS and NHSE are also leading to changes in the NHSE ILD Quality Dashboard. NHSE requires that specialist centres in England periodically submit ILD Quality Dashboard data, with data monitored nationally to ensure high quality services are available to patients in all parts of England. Going forward, completion of the UK IPF Registry will now also collect data that can be used to capture these data items (with data for patients with IPF being used as a proxy for the wider ILD service). Historically it has been difficult for centres to provide this data reliably, and use of the UK IPF Registry will ultimately make data collection easier and more robust.

As the BTS ILD Registry was launched over six years ago it was felt that a critical review of the datasets would be timely. This review took place in 2019, taking into account question response rates, the utility of collected data items and the resource constraints of participating sites. The datasets for the UK IPF and Sarcoidosis Registries have been shortened, reducing the time required for data entry.

BTS provides detailed data summaries biannually to sites participating in the BTS ILD Registry. These summaries include data from each of the UK IPF Registry and the UK Sarcoidosis Registry, with each site given summarised data for their own centre and also for the national dataset, enabling direct comparison between local and national figures. Centres are able to benchmark their services against national standards on an ongoing basis and generate annual audit data on their services. Audits for IPF care are currently recommended on three to five year cycles against the NICE clinical IPF guideline and IPF Quality Standards document[2] [3]. Registry data also provide detail on the current wait times patients are experiencing before being seen at specialist centres, and on the uptake of anti-fibrotic drug use.

In summary, it is well recognised that time pressures in the NHS remain high and resources are often stretched. It does take time to collect and submit data to registries, however the benefits can be immense. It is hoped, with UK IPF Registry data submission becoming part of the ILD service specification, that resources will in time be provided to help with this work. The BTS ILD Registry continues to try and capture the full impact of both IPF and sarcoidosis disease in the UK. It offers a platform to facilitate research and drive up standards of care for all patients. Accurate registry-derived epidemiological data will undoubtedly help with future planning of healthcare resources for these diseases.

Dr Lisa Spencer, Chair, BTS Lung Disease Registry Steering Group

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Patient registries play a pivotal role in understanding epidemiology, standards of care and long-term outcomes for patients. For the first six years of the BTS ILD Registry – comprising the UK IPF Registry and the UK Sarcoidosis Registry – the focus was on building a substantial base of high-quality data, and this can only increase as the UK IPF Registry will be mandated by NHSE for specialist centres from 2020.

The true impact of a clinical dataset is best realised when data are fully interrogated by researchers who are committed to improving patient care. To this end, BTS will be opening the ILD Registry datasets to requests for access from researchers from 2020.

Rebranding these registries to include UK *in their titles reflects their growth in size and impact since their launch in early 2013, and BTS remains committed to supporting this valuable resource. My sincere thanks to all involved in the production of this report, including the ILD Registry Steering Group and the Chair, Dr Lisa Spencer. A special thanks to all sites who have worked tirelessly to contribute data at a time when resources are strained – this Registry would not be possible without their input.*

Professor Jonathan Bennett
Chair, BTS Board of Trustees

BTS Lung Disease Registry Steering Group Membership 2019:

Dr Lisa Spencer, Chair
Dr Huzaifa Adamali, Consultant Respiratory Physician
Mr Howard Almond, Patient Representative
Dr Nazia Chaudhuri, Consultant Respiratory Physician
Dr Robina Coker, Consultant Respiratory Physician
Dr Wendy Funston, Trainee Respiratory Physician
Dr John Hutchinson, Consultant Respiratory Physician
Mr Steve Jones, Action for Pulmonary Fibrosis
Mrs Sarah Lines, ILD Specialist Nurse
Dr Philip Molyneaux, Consultant Respiratory Physician
Dr Lisa Nicol, Consultant Respiratory Physician
Mr Jack Richardson, SarcoidosisUK
Dr Katherine Spinks, Consultant Respiratory Physician

Miss Sally Welham, BTS Deputy Chief Executive
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.actionpulmonaryfibrosis.org/

SarcoidosisUK – www.sarcoidosisuk.org

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

Cover photograph: Malcolm Mason, a tireless ambassador for Action for Pulmonary Fibrosis, who sadly passed away in October 2018.

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, and of enabling and facilitating research which will lead to a greater understanding of their epidemiology and progression.

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

The Registry is open to all secondary care institutions in England, Scotland, Wales and Northern Ireland. At the end of October 2019, 64 sites across 54 Trusts/Health Boards had obtained approval to participate. A further 43 sites have the approval process underway. The current full list of participating sites is given on page 27.

Overall the ILD Registry includes over 2,950 patient records (2,474 IPF records and 497 sarcoidosis records).

Data Entry

Data entry for individual patient records is divided into three sections:

- Patient demographics information (age, gender, comorbidities, etc.).
- Clinical features on diagnosis and at current 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 published NICE IPF Quality Standards [3].

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 and 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 provide confirmation of approval to participate from the Caldicott Guardian before access is granted to allow Registry data entry.

All patient identifiable data (e.g. name, date of birth, postcode) is encrypted at the point of entry and visible only to Registry users in the centre that has entered the data (therefore identifiable data may only be accessed by members of 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 will vary based on a number of factors.

For example, the UK IPF Registry includes 2,474 patient demographics records. Data on smoking history is drawn from the demographics record, but the denominator for smoking history given on page 11 is 2,360. This is because there are only 2,360 records where smoking history was recorded in the UK IPF Registry. This is a 'real world' registry, so not all questions are fully completed by centres for every single patient.

- All denominators given in this report exclude cases where no response was recorded.

Additionally, the denominator for a particular data item may vary depending on whether responses where the clinician entered 'Not known' were included or excluded from the overall number of cases.

- Answer options of 'Not known' are excluded in this report unless otherwise stated.

Finally, 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 101%.

PART 1 – The UK Idiopathic Pulmonary Fibrosis Registry

Idiopathic pulmonary fibrosis (IPF) remains an incurable, progressive lung disease characterised by variable degrees of inflammation and scarring[4]. The symptom burden experienced by patients with IPF is high, with increasing symptoms of breathlessness, cough, fatigue and, in some individuals, anxiety and depression [5 6].

In a 2016 publication it was estimated that over 5,000 new IPF cases were diagnosed each year in the UK and over 30,000 people were living with the disease [7]. The disease prevalence is limited by a high mortality rate – median survival from diagnosis for IPF is only 3-4 years untreated [8]. IPF was expected to be the cause of at least 5,000 deaths in the UK in 2016 [7]. These outcomes are worse than for most forms of cancer [9]. They equated to ~1% of all UK deaths per year and represented ~4.6% of all respiratory disease related deaths. However, IPF mortality appears to be an ever-moving target. In July 2019, Navaratnam and Hubbard reported a new analysis showing that IPF is now causing closer to 7% of all UK respiratory deaths [10]. It is evident that IPF is a growing public health issue and the indicators are that this will continue. The incidence of IPF rises rapidly with age [11] and the UK population is ageing [12]. Published epidemiological studies estimating disease incidence and prevalence have been vital in highlighting IPF. However, going forward it is important we collect prospective data. The UK IPF Registry is well placed to help improve understanding of the true incidence and prevalence of IPF over time.

Despite the ongoing difficulties in the diagnosis and management of IPF much progress has been made in recent years. Robust international guidelines for IPF have been published and frequently updated, most recently in 2018 [4 13]. Undoubtedly these facilitated successful recruitment to the largest clinical trials in IPF to date [14-16]. Their positive outcomes have resulted in worldwide licensing of two anti-fibrotic therapies; pirfenidone (Esbriet™) and nintedanib (OFEV™). These drugs have provided hope for people with IPF. They slow disease progression, and evidence is emerging that they prolong survival [17-21]. However, the quest for improved therapies that halt or reverse IPF continues, as reflected in the increasing number of phase 1 to 3 clinical trials assessing new potential treatments [22].

People enrolled into clinical trials are often a pure cohort of individuals with a defined stage of disease and limited comorbidities, due to often very specific trial entry criteria. In the real world individuals with IPF are often older, are at various disease stages and have more comorbidities, as is evident in the data presented in this report. It is therefore reassuring to find that in real-world cohorts anti-fibrotic therapy is associated with prolonged survival, as indicated in data published, for example, from the Australian IPF Registry [23]. Registries can give a unique insight into the real-world impact of anti-fibrotic therapies on survival in IPF.

Specific to the UK, the development of the NICE guideline [2] on diagnosis and management of IPF, and the subsequent NICE quality standards [3], have laid the foundations for optimum delivery of patient-centred clinical care for IPF. It is important going forward that all hospitals caring for patients with IPF demonstrate that they are applying these standards. This has not always been the case in the recent past [24]. Contribution to the UK IPF Registry provides hospitals with reports so they can assess exactly how they are performing against these important national standards.

Inclusion criteria

Participating centres are requested to obtain written consent from and 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 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 31st October 2019

At the end of October 2019, 47 centres have contributed data to the UK IPF Registry (from the 64 sites participating in the wider BTS ILD Registry):

2,474 patient demographics records – an increase of 756 patients compared to October 2018

1,812 complete clinical/diagnosis records from the first clinic visit

1,402 follow up records representing 704 unique patients. These follow up records represent 39% (704/1,812) of all complete records, with patients where follow up data has been entered having a mean of two follow up records each.

1.1 DEMOGRAPHICS DATA

Demographics records are collected at the first clinic visit. The demographics data dashboard (Figure 1) is included on page 12.

Gender

Male	*79% (1,883/2,391)*
Female	*21% (508/2,391)*

Age

Mean age ± SD (yrs)	*73.5 ± 8.3*
Aged 60 – 69	*24% (576/2,358)*
Aged 70 and over	*70% (1,657/2,358)*

As expected for this disease group. Idiopathic pulmonary fibrosis is more common in males over the age of 65 [10].

Smoking history

Current	*4% (87/2,360)*
Ex	*66% (1,552/2,360)*
Never	*28% (669/2,360)*
Not known	*2% (52/2,360)*

IPF is a disease of unknown cause with possible risk factors including smoking. Over 90% of patients do not currently smoke.

Duration of chest symptoms

40% (941/2,338) of patients recorded on the Registry had chest symptoms for more than 24 months before their first clinic visit.

These data demonstrate that patients present late with significant symptom burden and disease severity. Earlier diagnosis of IPF remains a key challenge for healthcare professionals [25].

Comorbidities

The four most common comorbidities were hypertension (34%), ischaemic heart disease (21%), diabetes (20%) and symptoms of gastro-oesophageal reflux disease (18%).

A significant number of comorbidities were reported in this patient group who are mainly over the age of 60. Approximately 4,150 comorbidities were reported across 2,301 patients (12% – reported no comorbidities) on average giving 1.8 (SD±1.2) per patient with median 2 (IQR 1, 3), with a range of 0 to 7 per patient.

First degree relatives with IPF

Of the 1,996 records where family history was available, 6% (113) had at least one first degree relative with IPF.

Studies suggest up to 10% of patients with sporadic IPF may have an affected first degree relative.[26] Genetic intelligence around IPF continues to grow but it is still a research tool. The NHS does not yet offer genetic screening, due to insufficient data in the field. However, individual patient testing can be discussed with local genetics services.

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UK IPF Registry – Demographics Data

Summarised national data correct to 31/10/2019

Total number of records	2474

Mean age at presentation
73.5

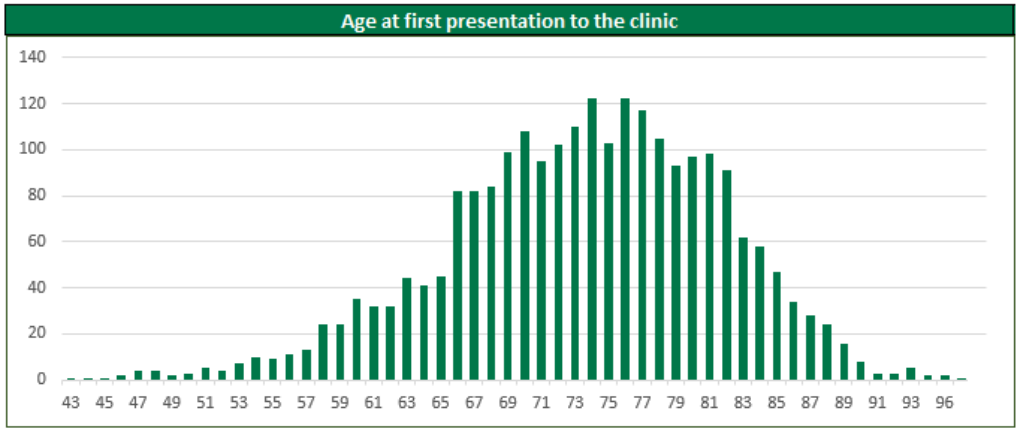
Mortality count
239

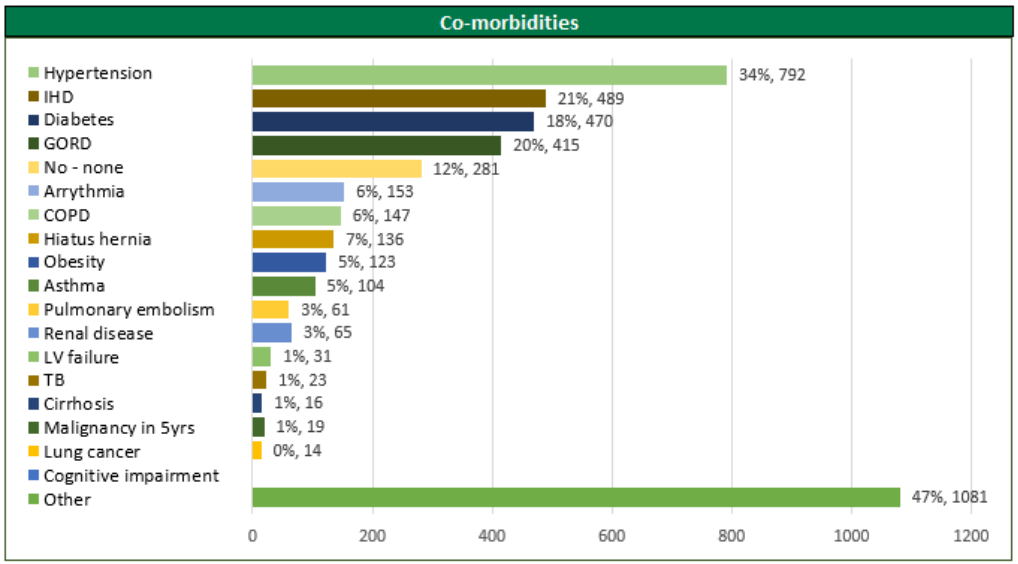
Mortality Rate
10%

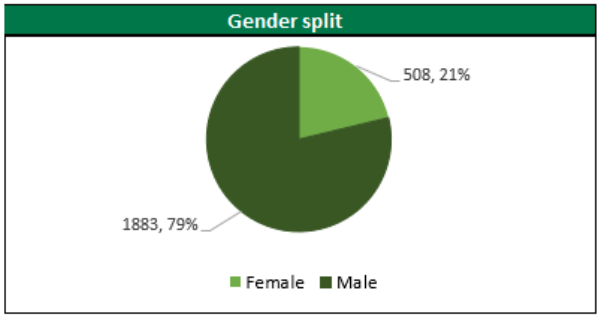
Symptom duration prior to presentation		
Duration	**Number**	**%**
Less than 6 months	216	10%
6-12 months	503	22%
12-24 months	541	24%
More than 24 months	941	42%
No symptoms	43	2%
Grand Total	**2244**	**100%**

First degree relatives with IPF		
Response	**Number**	**%**
Yes	125	7%
No	1765	93%
Grand Total	**1890**	**100%**

Smoking status		
Duration	**Number**	**%**
No never smoked/negligible	669	28%
Ex-smoker (quit >3m ago)	1552	66%
Current smoker	87	4%
Not known	52	2%
Grand Total	**2360**	**100%**







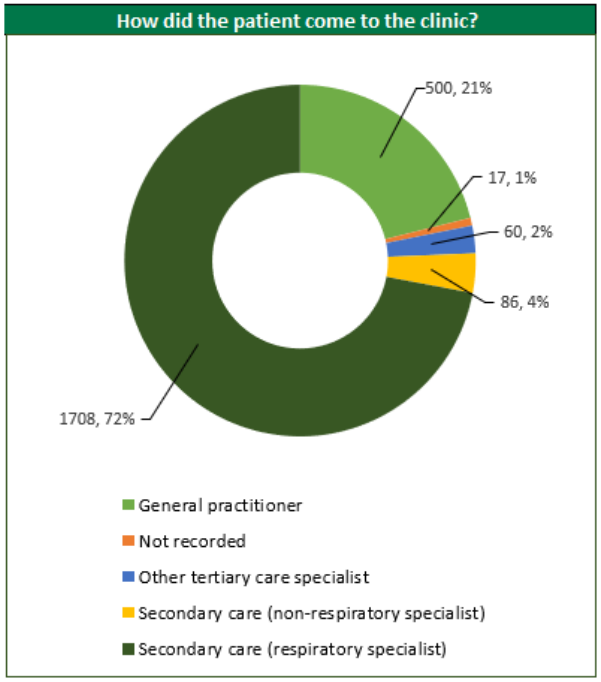


Figure 1: UK IPF Registry demographics data dashboard with figures correct to 31/10/2019

1.2 CLINICAL / DIAGNOSTIC DATA

Clinical/diagnostic records are collected at the first clinic visit. The clinical information data dashboard (Figure 2) is included on page 15.

Referral to first clinic visit (weeks)

10.9 weeks

(This is 11.2 weeks for specialist centres and 9.2 weeks for non-specialist centres.)

These wait times represent the time between referral receipt and the first clinic visit. Therefore, they do not represent referral to treat times (RTT) for all hospitals represented in these data. Some sites conduct a virtual ILD MDT before first clinic attendance, some conduct their MDT the same day as the first clinic visit, some conduct their ILD MDT after first clinic visit and may need to bring the patient back to hit an RTT NHS treatment target point.

If patients are first seen in their local centre then are referred to a specialist centre the wait to confirm a diagnosis or start treatment can be over 5 months.

It would be beneficial for patients if these waits could be reduced going forward, particularly given the poor prognosis of untreated IPF. The Registry permits monitoring of waits

Lung Function (at presentation)

FVC predicted >80%	*38% (686/1,809)*
FVC predicted 50-80%	*57% (1,027/1,809)*
FVC predicted < 50%	*5% (96/1,809)*

At entry 38% of patients are above current NICE criteria for anti-fibrotic treatment. However, Registry data show these patients already exhibit a substantial decrease in their diffusion capacity (mean TLCO= 43%) and only a small percentage of these patients (7%) have significant radiological emphysema.

These figures require careful interpretation. The majority of records were entered from ILD specialist centres, with IPF populations naturally skewed towards those within the anti-fibrotic treatment range defined by NICE (50-80% of predicted FVC) or those that may move into that range from higher FVC values. However, it does suggest that a significant proportion of patients with IPF cannot access anti-fibrotic drugs at the point of diagnosis due to their FVC being above 80% predicted.

Patients with FVC values <50% predicted are less likely to be referred on to a specialist centres, as treatment cannot be accessed and/or they may be too unwell to consider travel. This likely explains why only 5% of patients on the UK IPF Registry have lower FVC values.

Surgical lung biopsy rate

Yes	*8% (146/1,809)*
No	*89% (1,602/1,809)*
Not known	*3% (61/1,809)*

Previous Registry data demonstrated a UK lung biopsy rate in IPF of 13% [27], which appears to be falling. When figures are split by year of presentation, Registry data show a fall in surgical lung biopsy rates from 16.1% in 2013 to 1.2% in 2019.

This is consistent with published data [28]. New IPF international diagnostic guidelines published in 2018 [4 13] now make it possible for more patients to secure a firm diagnosis of IPF without the need for lung biopsy. This is helpful as many patients are unsuitable to undergo lung biopsies due to their age and comorbidities. More patients can therefore access drug treatment for their IPF.

Current drug treatment

Received pirfenidone	*26% (457/1,750)*
Received nintedanib	*19% (339/1,750)*
No anti-fibrotic treatment	*45% (790/1,750)*

These data need to be interpreted carefully. They reflect drug use over the whole timescale of the Registry from 2013 onwards. Pirfenidone has been available through the NHS since 2013, whereas nintedanib has only been available since 2016. Some patients on the IPF Registry may have had access to drugs through pharma-supported programmes before their NICE approval was granted, with eligibility criteria different to that approved through NICE.

Suitability for drug treatment is not based purely on FVC criteria. Other factors, such as renal or liver function abnormalities, can sometimes preclude use of drugs. Further analysis of UK IPF Registry data (not presented here) can reveal additional information regarding drug treatment.

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UK IPF Registry – Clinical Information Data

Summarised national data correct to 31/10/2019

Count of Records
1812

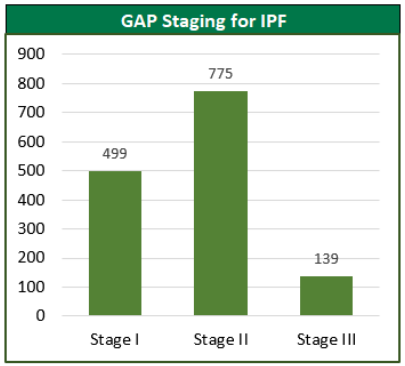
Referral to 1st clinic visit (wks)
10.9

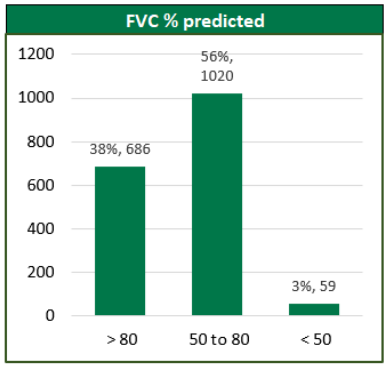
HRCT		
HRCT pattern	Number	%
Definite UIP	758	44%
Possible UIP	855	50%
Inconsistent	77	4%
Not known	32	2%
Grand Total	1722	100%

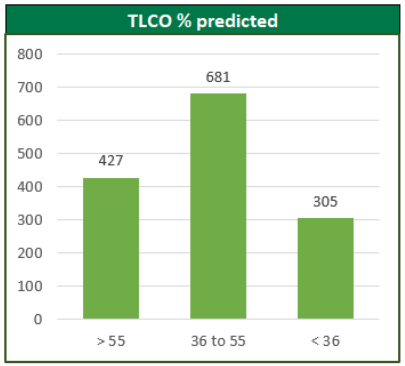
Surgical biopsy		
Biopsy taken?	Number	%
No	1602	89%
Yes	146	8%
Not known	61	3%
Grand Total	1809	100%

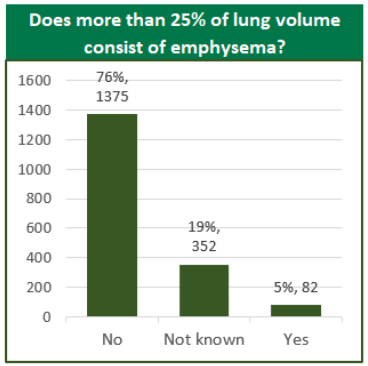
Lung transplant		
Patient referred?	Number	%
Yes	53	3%
N/A at this time	482	27%
N/A at any time	1142	63%
Not known	132	7%
Grand Total	1809	100%

Clinical trials		
Patient recruited?	Number	%
Yes	87	5%
No	1532	86%
Not known	169	9%
Grand Total	1788	100%







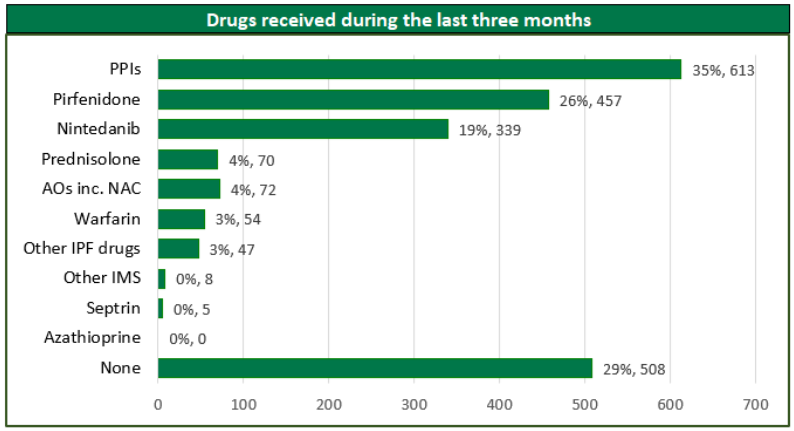


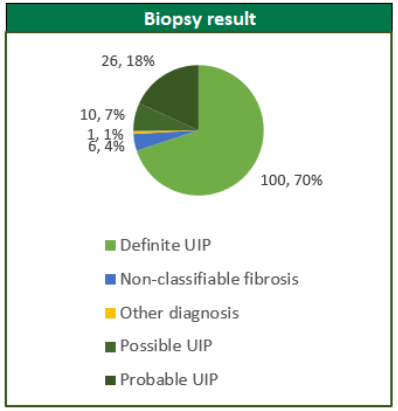
6MWT on air - mean distance	
284.8	metres
6MWT on air	standard deviation
164.0	metres

Ability to perform 6MWT on air		
Is patient able?	Number	%
Yes	1315	88%
No	187	12%
Grand Total	1502	100%

6MWT Performance		
Patient did 6MWT	Number	%
Yes	638	53%
No	574	47%
Grand Total	1212	100%

Discussion at MDT		
MDT discussion	Number	%
Yes	1606	91%
No	84	5%
Pending	78	4%
Grand Total	1768	100%





Is the patient on oxygen?		
Oxygen	Number	%
Yes: short burst	15	1%
Yes: LTOT	101	5%
Yes: ambulatory	260	14%
Yes: palliative	1	0%
No	1469	80%
Grand Total	1846	N/A

Pulmonary Rehabilitation		
Patient needs assessed?	Number	%
Yes, and referred	444	59%
Yes, but had PR in last 12m	47	6%
Yes, but NOT SUITABLE	132	18%
Yes, patient declined	79	11%
No, not assessed	46	6%
Grand Total	748	100%

Palliative care		
Patient assessed?	Number	%
Yes	590	62%
No	131	14%
Not known	238	25%
Grand Total	959	100%

Figure 2: UK IPF Registry clinical information/diagnosis data dashboard with figures correct to 31/10/2019

1.3 FOLLOW UP DATA

Follow up information is requested every 12 months after the first clinic visit. However, clinicians may upload follow up data from any clinic visits at any time, regardless of timescale. All follow up records are gratefully received and are analysed fully. Records may be interrogated based on length of time (in months) since the first clinic visit. The follow up data dashboard (Figure 3) is included on page 17. Only selected facts are presented here.

Lung cancer reported rate

2% (15/704) of patients at follow up were diagnosed with lung cancer since last review.

(Follow up periods range from six months to 6.5 years.)

This figure is likely an underestimate due to the limited availability of follow up data within the UK IPF Registry (only 28% of the overall 2,474 patients have at least one follow up record).

Figures for national incidence of lung cancer by age and sex are available from Cancer Research UK [9].

Hospital admissions

12% (81/704) of cases had at least one acute hospital admission with respiratory disease following initial presentation. Of these:

23% (19/81) included at least one acute exacerbation of IPF;

51% (41/81) included at least one admission for pneumonia/ respiratory tract infection.

To date only 28% of incident IPF cases have follow up records included. The hospitalisation rate here needs to be considered within this context. As the number of follow up records increases the reliability of these data will improve.

Death since entry onto Registry:

10% (239/2,474)

Of these, acute or chronic progression of IPF was the cause of death in 78 cases (46% of those cases where cause of death was recorded).

This dataset covers a period of 6.5 years maximum and is incomplete at this time. This recorded mortality rate is much lower than the annual mortality expected in a generic IPF population. We therefore expect the true burden of disease to be shown as the number of follow up records in the UK IPF Registry increases.

British Thoracic Society

UK IPF Registry – Follow Up Data

Summarised national data correct to 31/10/2019

Count of Records
1402

Unique patients
704

HRCT since last review?		
HRCT	Number	%
Yes	216	15%
No	1179	84%
Not known	4	0%
Grand Total	1399	100%

Most recent HRCT pattern		
HRCT pattrn	Number	%
Possible UIP	73	37%
Definite UIP	109	55%
Inconsistent	9	5%
Not known	8	4%
Grand Total	199	100%

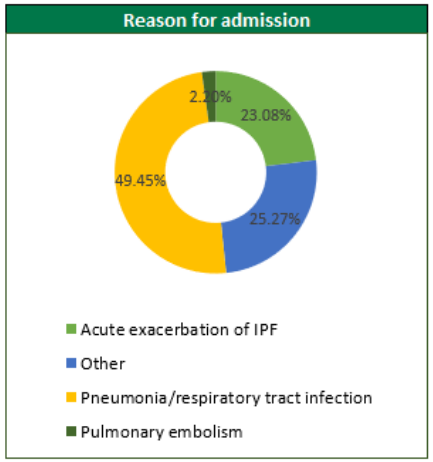
Acute hospital admissions - respiratory		
Since presentation	Number	%
Yes	96	7%
No	1154	86%
Not known	85	6%
Grand Total	1335	100%

Referral for transplant		
Patient referred	Number	%
Yes	61	9%
N/A at this time	381	55%
N/A at any time	191	27%
Not known	64	9%
Grand Total	697	100%

Is the patient on oxygen?		
Oxygen therapy	Number	%
Yes - LTOT	86	6%
Yes - ambulatory	225	16%
Yes - both	0	0%
No	1058	75%
Not known	27	2%
	N/A	N/A

Patient reassessed for oxygen?		
Patient referred	Number	%
Yes	380	40%
No	151	16%
Not known	418	44%
Grand Total	949	100%

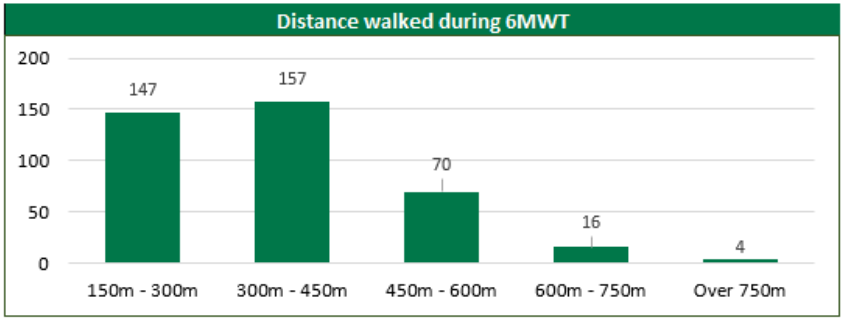
Reason for admission



6MWT on air		
Is patient able?	Number	%
Yes	828	59%
No	146	10%
Not known	425	30%
Grand Total	1399	100%

6MWT on air		
Is patient able?	Number	%
Yes	411	42%
No	570	58%
Grand Total	981	100%

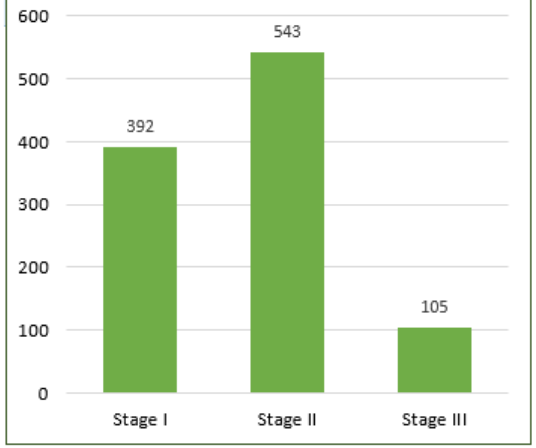
Distance walked during 6MWT



Average of oxygen saturation at rest (%)
95.7 %

Average lowest O2 saturation in walk (%)
87.5 %

GAP Staging for IPF



Drugs received during the last three months

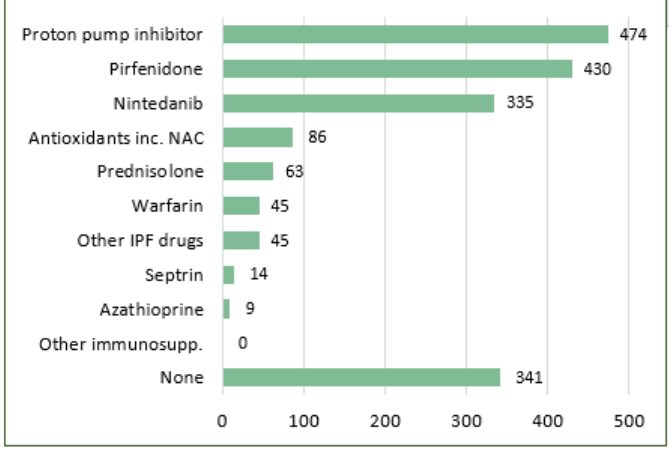


Figure 3: UK IPF Registry follow up data dashboard with figures correct to 31/10/2019

1.4 HOW DATA COLLECTED THROUGH THE UK IPF REGISTRY RELATE TO NICE QUALITY STANDARDS FOR IPF

In this section, data from the UK IPF Registry are presented in relation to each published NICE Quality Standard [3]. The initial UK IPF Registry dataset was designed before the publication of the NICE Quality Standards for IPF, so not all data points are yet available. Data suggest there is room for improvement in general in UK clinics to ensure patients with IPF are always treated according to these standards. The UK IPF Registry can assist with assessing improvement in the delivery of patient care as measured by these standards.

IPF Quality Statement	Registry data	Comments
Quality Statement 1: **People are diagnosed with IPF only with the consensus of a multidisciplinary team (MDT) with expertise in interstitial lung disease.**	89% (1,606/1,809) of cases were discussed at an ILD MDT, whereas 5% (84/1,809) were not discussed Registry data indicate the mean time from patient referral to MDT was 10.7 weeks (10.6 weeks for specialist centres and 11.4 weeks for non-specialist centres). Each of these figures had a high standard deviation (±14 to 18 weeks) indicating some MDTs may be held before or after the first clinic appointment.	Note: 119/1,809 case records showed that MDT was pending or 'not known' at data entry. The target for this standard would ideally be 100%.
Quality Statement 2: **People with IPF have an interstitial lung disease specialist nurse available to them.**	The current UK IPF Registry dataset does not capture information regarding the provision of ILD specialist nurses in centres, or whether patients have an ILD specialist nurse available to them. To address this important quality statement a new question is being added to the UK IPF Registry. This new question will identify interactions with ILD nurses on a patient by patient basis. UK IPF Registry publications will report on this going forward.	
Quality Statement 3: **People 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.**	At presentation, Registry data show that 18% (321/1,790) of patients are on oxygen (at least one of ambulatory, LTOT, short burst or palliative). Assessment of long-term oxygen needs takes place for 95% (896/944) of patients at presentation and is recorded in 95% (804/848) of follow up records.	The target for assessment of LTOT would ideally be 100% to hit the standard. The current UK IPF Registry dataset does not accurately capture information regarding assessment of ambulatory oxygen. However, this information will be collected going forward.

Quality Statement 4: **Pulmonary rehabilitation programmes provide services that are designed specifically for IPF.**	Patient assessment and referral rates for pulmonary rehabilitation (PR) at presentation show: 74% of patients (702/951) were assessed for PR; 5% (46/951) of patients were not (informtation was not recorded for the remaining cases). Of those assessed, 63% (444/702) were referred for PR. Where patients were assessed but not referred the reasons were: - 18% (47/258) had attended PR within the previous 12 months; - 51% (132/258) were not suitable due to poor mobility or a very good fitness level; - 31% (79/258) of patients declined.	The Registry has not collected data on whether PR programmes provided are designed specifically for patients with IPF. There remain parts of the UK where PR cannot be accessed for patients with IPF or other ILDs, as services were originally set up and funded for patients with COPD. Published data have shown PR programmes can be very beneficial for patients with IPF despite the progressive nature of the disease [29]. Going forward the NHS long-term plan may help to address these geographical inequalities.
Quality Statement 5: People with IPF and their families and carers have access to services that meet their palliative care needs.	Data for palliative care considerations at first clinic visit show 62% (590/959) of patients had their palliative needs assessed and managed, while 14% (131/959) of patients did not (remaining cases were recorded as 'not known'). For follow up visits these figures have reduced, with palliative care needs being assessed and managed at only 40% (380/949) of visits, and not in 16% (151/949) of visits (remaining cases were recorded as 'not known').	These low figures may in part reflect a misunderstanding of the UK IPF Registry question *Have you assessed and managed the palliative care needs of this patient at this clinic visit?* When asked why palliative care needs had not been assessed and managed, some clinicians indicated the patient had no current needs. The UK IPF Registry question is being amended to ensure clinicians are aware that identifying a patient has no current palliative needs is still conducting an assessment to ensure patient needs are met. Symptom management of patients with IPF remains a critical part of overall care. Published studies have shown that symptom burden in patients with IPF is high [5, 6].

PART 2 – The UK Sarcoidosis Registry

Sarcoidosis is a disease with an incidence of around 7 per 100,000 per year in the UK [30]. 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. While genetic and environmental factors may play a role in whether someone develops sarcoidosis, the exact cause of the disease is unknown [31].

The presentation of sarcoidosis, including organs affected and severity, may be linked to the age, sex and ethnicity of the patient.[32] Epidemiological data suggest sarcoidosis is most prevalent in Northern European and African-American populations (the latter having higher severity disease), with lower rates reported in Japan. It typically occurs in patients between 20 and 50 years of age, but can occur in those over 60, particularly in women. Some studies report similar prevalence between genders, while others report higher rates in women [30, 32-34].

Some patients with sarcoidosis are diagnosed incidentally on imaging and remain asymptomatic, while others have significant symptoms. Some patients will present acutely but later enter remission, while others – up to 30% – have a more chronic, progressive course. Many patients are limited by breathlessness, cough, fatigue or joint pains, with subsequent impact on quality of life and ability to work or fulfil care responsibilities.

Management of sarcoidosis includes education, monitoring, supportive care and, in certain patients, immunosuppression. Commonly used agents include oral corticosteroids and methotrexate. Some patients may benefit from biologic therapies such as infliximab. Patients may also be offered oxygen therapy, pulmonary rehabilitation and referral for lung transplantation. Around 1-5% of patients with sarcoidosis die from complications of their disease, commonly progressive respiratory failure or cardiac disease [35].

The British Lung Foundation's *Battle for Breath* report indicates that sarcoidosis resulted in nearly 9,000 hospital bed days in the UK in 2011.[7] According to the Department of Health, the average bed day is estimated to cost £400, and therefore hospital admissions for sarcoidosis can be estimated to cost in the region of £3.5 million per annum. North American data suggest that those with more complicated sarcoidosis can have considerably higher healthcare needs [34].

Since its inception, the UK Sarcoidosis Registry has amassed over 490 cases from across the UK. The hope is that these cases will contribute to a greater understanding of the characteristics of people with sarcoidosis across the country, ultimately leading to earlier recognition and improvements in diagnosis and management.

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 31st October 2019

At the end of October 2019, 34 centres have contributed data to the UK Sarcoidosis Registry (from the 64 sites participating in the wider BTS ILD Registry):

497 patient demographics records

357 complete clinical/diagnosis records from the first clinic visit

143 follow-up records representing 81 unique patients.

2.1 DEMOGRAPHICS DATA

Demographics records are collected at the first clinic visit. The demographics data dashboard (Figure 4) is included on page 23.

Gender

Female	*42% (197/464)*
Male	*58% (267/464)*

Although many studies note a higher proportion of females, recent data from the British Lung Foundation suggest no difference in incidence between males and females, based on primary care data [30].

Age

The age distribution is shown in Figure 4. Mean age at presentation was 49.8 years, with a standard deviation of ± 13.2 years.

A significant number of patients in the UK Sarcoidosis Registry presented over the age of 50, with some over 80. This suggests the diagnosis should be considered even in older patients. A recent North American database study supports this, with over half of patients aged over 55 at diagnosis [34].

Ethnicity

67% (294/437) were White British.

Sarcoidosis in known to be more prevalent in black populations. This figure likely reflects the populations from which Registry data were obtained. According to UK census data from 2011, 86% of the population of England and Wales reported their ethnicity as White, whereas only 3% described themselves as Black / African / Caribbean / Black British [36].

Referral

50% (224/451) of patients were referred from respiratory physicians in secondary care.

This likely reflects the complexities in diagnosing sarcoidosis and the increasing sub-specialism in respiratory medicine. Lack of awareness in primary care may play a part, but referral pathways could also be streamlined so patients are seen by the appropriate person as soon as possible. These factors could lead to delay in diagnosis, although this proportion is lower than in previous years.

Smoking history

9% (28/324) of patients were smokers at presentation and 27% (88/324) were ex-smokers.

This is a lower proportion than in the wider population, where 16.8% of 45-54 year olds (encompassing the mean age of diagnosis in the Registry) were smokers [37]. The numbers are too small to draw firm conclusions, but may reflect earlier, successful attempts at smoking cessation at the time of symptom onset.

Comorbidities

Over one third of patients (39%, 127/324) had no comorbidity at the time of their current presentation.

The most common comorbidities highlighted were systemic hypertension (15%), diabetes (11%), asthma (9%) and obesity (6%). These conditions are highly prevalent in the general population, and should be taken into account when considering treatment initiation and predicting impact in individual patients – for example, the risks of corticosteroids contributing to weight gain and impaired glycaemic control.

British Thoracic Society

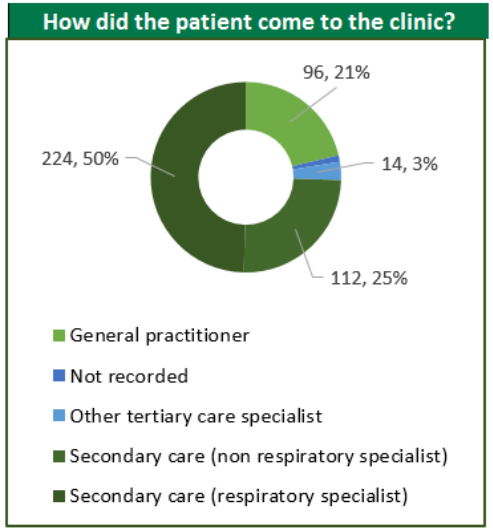
UK Sarcoidosis Registry – Demographics Data

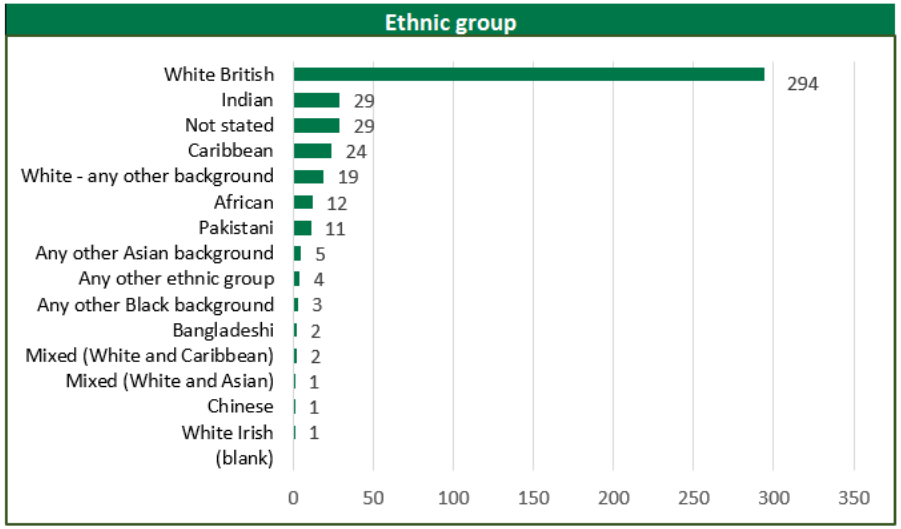
Summarised national data correct to 31/10/2019

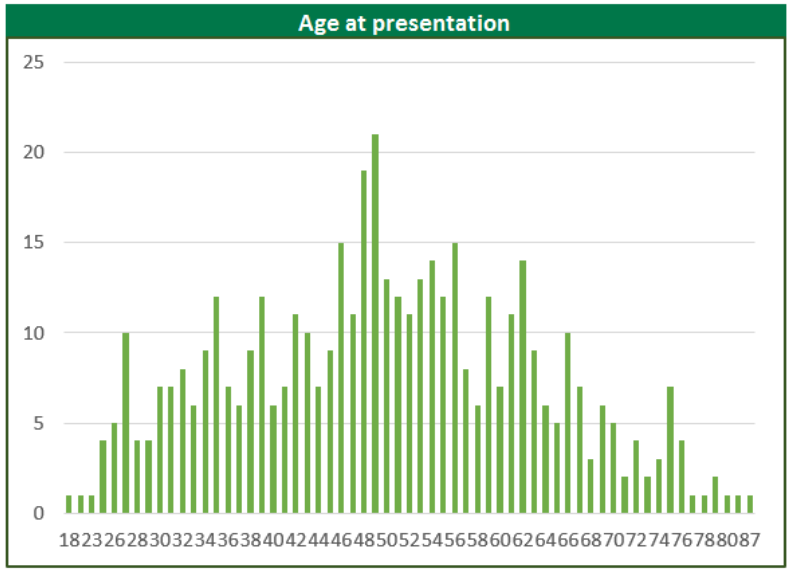
Total number of records
497

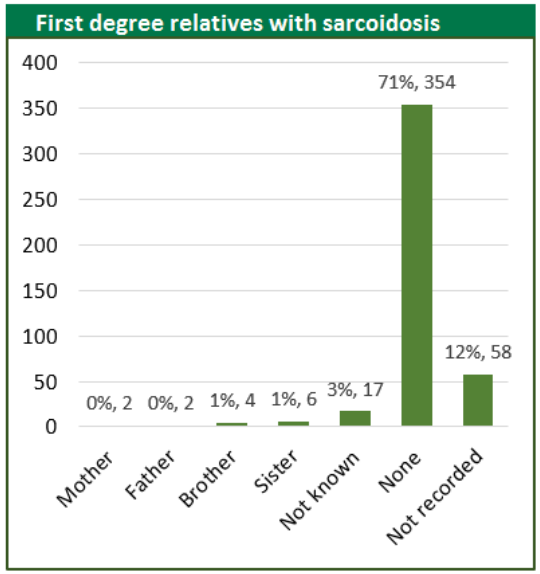
Mean age at presentation
49.8

Smoking status		
At presentation	Number	%
Current smoker	28	9%
Ex-smoker	88	27%
Never smoked	199	61%
Unknown	9	3%
Grand Total	324	100%









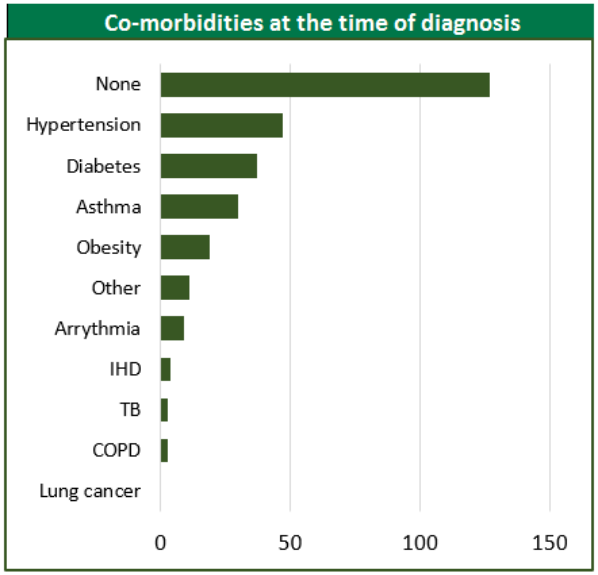


Figure 4: UK Sarcoidosis Registry demographics data dashboard with figures correct to 31/10/2019

2.2 CLINICAL / DIAGNOSTIC DATA

Clinical/diagnostic records are collected at the first clinic visit. The clinical information data dashboard (Figure 5) is included on page 26.

Symptoms at current presentation

The most common symptoms were breathlessness (52% - 167/322) and cough (45% - 145/322).

Fatigue (30%), joint pain (15%) and eye symptoms (13%) were also frequently reported.

Treatment of fatigue in sarcoidosis is challenging, and better therapeutic options are needed. The high burden of fatigue in the Registry highlights this as an unmet need and a target for future research – both pharmaceutical and more holistic options, for example pulmonary rehabilitation could be considered [38].

Biopsy

Over two thirds (70%) of diagnoses were confirmed through biopsy.

Endobronchial ultrasound (EBUS) was used in 28% of biopsies in 2013, rising to 43% in 2019 (overtaking mediastinoscopy and transbronchial biopsy as the most common biopsy techniques for confirming pulmonary disease).

Incidental diagnosis

In one third of patients (35%, 111/318) diagnosis was the result of an incidental finding during the course of investigations for another condition.

This highlights the importance of wider education of healthcare professionals about sarcoidosis. Increasing incidental detection of sarcoidosis due to imaging for other reasons may lead to an increased incidence of disease but lower proportion of those with severe symptoms.

Blood test

The most common finding from blood tests was peripheral lymphopaenia, identified in over half of patients (57%, 135/237) at presentation.

Although non-specific, peripheral lymphopaenia should raise the possibility of sarcoidosis within the correct clinical context.

Patients presented with mean serum IgG of 15.6g/L (ranging from 2.5g/L to 67g/L). 29% of patients had IgG levels above the normal range of 5-16g/L.

There has been interest in using immunoglobulin levels as a disease marker in sarcoidosis, although recent data suggest no association between levels at diagnosis and evolution of disease [39].

The mean ACE level at presentation was 67.6 IU/L with 51% of patients having an ACE level in excess of 55 IU/L. At follow up the mean ACE level dropped to 54.0.

Serum ACE levels are widely used as a disease marker for sarcoidosis, despite limitations, although guidelines do not recommend using serum ACE for diagnosis.

Current treatment

Most, patients were either not started on treatment (42%), or managed with systemic corticosteroids (51%). Alternative agents varied with no preponderance of any one agent.

This broadly reflects previous BTS guidance on the management of sarcoidosis [40].

Inclusion in clinical trials

While only one patient was known to be recruited into a clinical trial, 94% (276/294) said they would like to be considered for recruitment subject to inclusion criteria.

There is a need for more clinical research in sarcoidosis. The Registry may provide a cohort of interested and suitable patients. Current questions include optimal time to initiate treatment, dosing regimens, the role of alternative immuno-suppression, treatment duration, and management of associated symptoms such as fatigue.

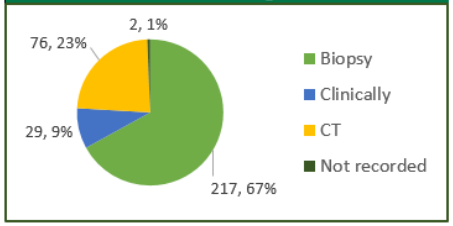
British Thoracic Society

UK Sarcoidosis Registry – Clinical Information Data

Summarised national data correct to 31/10/2019

Total number of records
357

Method of diagnosis



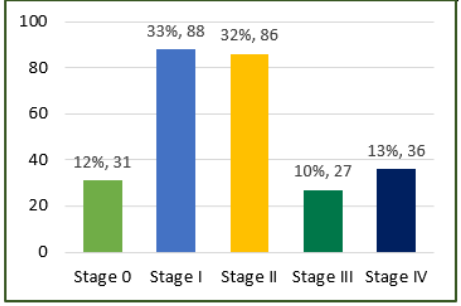
Bronchoscopy

Bronchoscopy	Number	%
Yes	109	36%
No	187	62%
Unknown	7	2%
Grand Total	303	100%

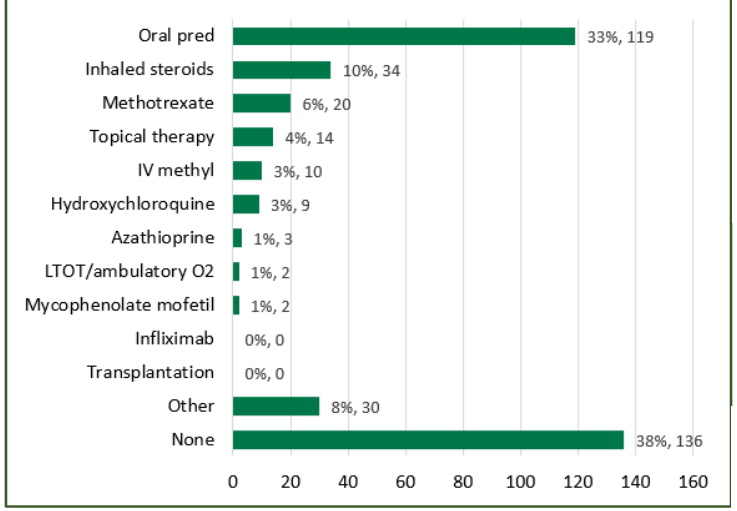
Bronchalveolar lavage

Bronchoalveolar lavage	Number	%
Yes	41	13.5%
No	254	83.8%
Unknown	8	2.6%
Grand Total	303	100%

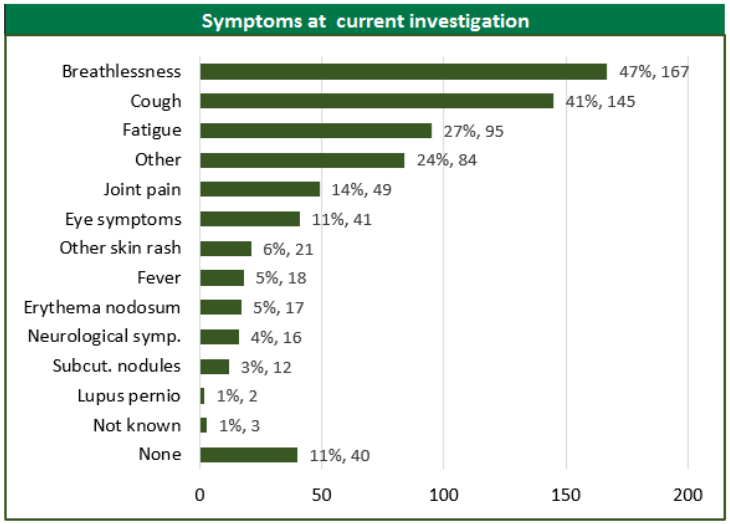
Sarcoidosis stage from chest radiograph



Treatment record at current investigation



Symptoms at current investigation



Was diagnosis incidental?

Incidental?	Number	%
Yes - radiology	84	26%
Yes - blood tests	5	2%
Yes - skin biopsy	5	2%
Yes - other	17	5%
No	203	64%
Not recorded	4	1%
Grand Total	318	100%

Was sarcoidosis confirmed on histology?

Histology	Number	%
Yes	228	70%
No	93	29%
Not recorded	3	1%
Grand Total	324	100%

Biopsy site (surgical and non-surgical)

Biopsy site	Number	%
Lung	204	57.1%
Liver	4	1.1%
Skin	17	4.8%
Extrathoracic LN	24	6.7%
Brain	0	0.0%
Heart	2	0.6%
Kidney	0	0.0%
Other	11	3.1%
Grand Total	N/A	N/A

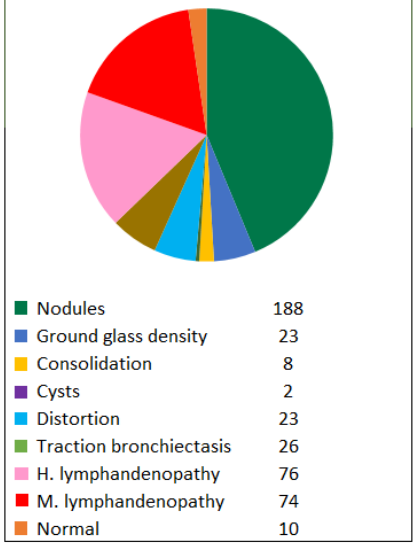
MRC dyspnoea grade

Dyspnoea grade	Number	%
Grade 1	143	46%
Grade 2	88	28%
Grade 3	29	9%
Grade 4	13	4%
Grade 5	2	1%
Patient not breathless	22	7%
Not recorded	16	5%
Grand Total	313	100%

Average of	ACE level
67.57	µg/l

Average of Immunoglobulin G (IgG) g/l
14.56 g/l

Most recent HRCT



Most recent blood test results

Results	Number	%
Lymphopenia	135	62%
Raised ESR	39	18%
Raised CRP	41	19%
Abn. liver function	48	22%
Raised Ca2++	22	10%
Other abnormality	46	21%
Grand Total	N/A	N/A

Figure 5: UK Sarcoidosis Registry clinical information/diagnosis data dashboard with figures correct to 31/10/2019

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
Birmingham Heartlands Hospital, Heart of England NHS Trust
Castle Hill Hospital, Hull and East Yorkshire Hospitals NHS Trust
Central Middlesex Hospital, London North West Healthcare NHS Foundation Trust
Cheltenham General Hospital, Gloucestershire Hospitals NHS Foundation Trust
Churchill Hospital, Oxford University Hospitals NHS Trust
City Hospital, Sandwell and West Birmingham NHS Trust
Countess of Chester Hospital, Countess of Chester Hospital NHS Foundation Trust
Croydon University City Hospital, Croydon Health Services NHS Trust
Darlington Memorial Hospital, County Durham and Darlington NHS Foundation Trust
Ealing Hospital, London North West Healthcare NHS Foundation Trust
George Eliot Hospital, George Eliot Hospital NHS Trust
Glenfield Hospital, University Hospitals of Leicester
Gloucestershire Royal Hospital, Gloucestershire Hospitals NHS Foundation Trust
Good Hope Hospital, Heart of England NHS 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
Hinchingbrooke Hospital, Hinchingbrooke Health Care NHS 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, Taunton & Somerset NHS Foundation Trust
New Cross Hospital, Royal Wolverhampton Hospitals 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
Northwick Park Hospital, London North West Healthcare NHS Foundation Trust
Nottingham City Hospital, Nottingham University Hospitals NHS Trust
Papworth Hospital, Papworth Hospital NHS Foundation Trust
Peterborough City Hospital, Peterborough & Stamford Hospitals NHS Foundation Trust
Queen Alexandra Hospital, Portsmouth Hospitals NHS Trust
Queen Elizabeth Hospital, Gateshead Health NHS Foundation 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 Victoria Infirmary, Newcastle upon Tyne Hospitals NHS Foundation Trust
Russells Hall Hospital, The Dudley Group NHS Foundation Trust
Solihull Hospital, Heart of England NHS Trust
Southampton General Hospital, University Hospital Southampton NHS Foundation Trust
Southmead Hospital, North Bristol NHS Trust
St James' University Hospital, 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 Aintree, Aintree University Hospitals NHS Foundation 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
Glasgow Royal Infirmary, NHS Greater Glasgow and Clyde
Lorn & Islands District General Hospital, NHS Greater Glasgow and Clyde
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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