BMJ Open Respiratory Research

Idiopathic pulmonary fibrosis in the UK: findings from the British Thoracic Society UK Idiopathic Pulmonary Fibrosis Registry

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To cite: Fahim A, Loughenbury M, Stewart I, *et al*. Idiopathic pulmonary fibrosis in the UK: findings from the British Thoracic Society UK Idiopathic Pulmonary Fibrosis Registry. *BMJ Open Respir Res* 2025;**12**:e002773. doi:10.1136/bmjresp-2024-002773

► Additional supplemental material is published online only. To view, please visit the journal online (https://doi.org/10.1136/bmjresp-2024-002773).

Received 29 July 2024
Accepted 7 January 2025

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ABSTRACT

Objectives Idiopathic pulmonary fibrosis (IPF) is a progressive interstitial lung disease (ILD) and the most common idiopathic interstitial pneumonia. The UK IPF Registry was established in 2013 to collect data pertaining to clinical features, therapeutic approaches and outcomes. From February 2023, the Registry expanded to include any ILD with evidence of fibrosis.

Design The UK IPF Registry is a national, multicentre observational registry, including both prospective and retrospective data of patients with IPF in secondary or tertiary care. Cases eligible for inclusion were those with a diagnosis of IPF, presenting at participating centres from January 2013.

Results Between January 2013 and February 2023, 5052 IPF cases were registered from 64 participating centres. There was a male preponderance (77.8%) with mean±SD age of 74±8.1 years, 66% were ex-smokers and 76% had at least one comorbidity. Over a third (36.7%) experienced symptoms for more than 24 months prior to their first clinic visit. The majority of cases were discussed at a multidisciplinary team (MDT) meeting and the most common radiological patterns at presentation were probable (54.6%) and definite (42.7%) usual interstitial pneumonia. There was a reduction in surgical lung biopsies from 14% in 2013 to 5.5% in 2022. Antifibrotic therapy prescription rose from 36.0% in 2013 to 55.9% in 2023. The use of nintedanib (approved by National Institute of Clinical Excellence in January 2016) rose from 6.7% in 2013 to 31.5% in 2022 and pirfenidone (approved in April 2013) was initially used in around a third of cases before dropping to between 16.8% and 24.9% after nintedanib was approved.

Conclusion These data reflect clinical practice across the UK and it is intended the data will have a role in informing the future of IPF care and providing a model for benchmarking, ultimately increasing knowledge and improving clinical care for this devastating disease.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Idiopathic pulmonary fibrosis (IPF) is a progressive fibrotic disorder with a dismal prognosis and there are several data sets/registries, collecting information on the clinical behaviour and outcomes of this disease. However, there is no long-term published data from the UK IPF registry that would inform clinical practice in the UK.

WHAT THIS STUDY ADDS

⇒ The findings of this evaluation provide real-world IPF data and describe the aspects of diagnosis and management of this disease over a decade. It shows the impact of changes in guidelines on clinical practice with reductions in lung biopsies and an increase in antifibrotic therapy over time.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ The findings of this largest single-country IPF registry highlight key areas to be addressed in providing appropriate clinical care to this group of patients in the UK. It is imperative that appropriate National Health Service resources and funding are made available for patients with IPF, including earlier access to antifibrotic therapy and recruitment into clinical trials, with the aim of improving clinical outcomes and quality of life for patients with IPF and their caregivers.

INTRODUCTION

Idiopathic pulmonary fibrosis (IPF) is a progressive interstitial lung disease (ILD) with a significant symptom burden and dismal prognosis.[1] Patients present with breathlessness, cough and fatigue. IPF impacts quality of life, physically, emotionally, psychologically and socially.[2] Progression varies and we cannot accurately identify the cohorts likely to progress more rapidly.

Registries are important tools to enhance knowledge and learning in long-term conditions such as IPF as they provide 'real world' information about disease behaviour,

treatment patterns and a range of other variables and are not restricted to the limiting entry criteria of clinical trials. Furthermore, disease registries can be invaluable in capturing longitudinal data over a much longer period than randomised controlled trials.

The UK IPF Registry was established to increase real-world knowledge of IPF in the UK and to help improve management and service delivery. We present data collected over the first decade from 2013 to 2023.

METHODS

Registry design and participation

The UK IPF Registry (established in 2013) is a national, multicentre, observational registry of prospective and retrospective IPF cases across the UK.[3] Data are entered via a purpose-built web-based platform (https://registry.brit-thoracic.org.uk/) by participating hospital teams. The service is funded by British Thoracic Society (BTS) and grants were previously received from the Healthcare Quality Improvement Partnership, Boehringer Ingelheim and InterMune (2011–2014).

Patient and public involvement

Patient and public representatives were involved in the development of the BTS Lung Disease Registry Programme, through the patient charities Asthma+Lung UK and Sarcoidosis UK and the lay member of the BTS Board of Trustees. Patient perspectives on governance, design and communication are embedded into the programme through ongoing representation from the patient charities Action for Pulmonary Fibrosis and Sarcoidosis UK as well as oversight from the BTS Board.

Inclusion criteria

The inclusion criteria are: patients with definite or strongly suspected IPF, who have either:

- A new diagnosis of IPF made at a clinic visit from 1 January 2013, or
- A historical diagnosis of IPF, was seen for the first time in the clinic at the participating centre from 1 January 2013.

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

Data collection

Baseline demographic, diagnosis, clinical features and treatment data were collected at the first clinic visit, with follow-up information requested every 12 months. The data set for the initial outpatient appointment is in online supplement 1 and the follow-up is in online supplement 2.

Patient-identifiable data are encrypted on entry. Data may be entered retrospectively, provided the first clinic visit was on or after 1 January 2013.

Data management

Participating sites entered data directly onto the purpose-built secure platform. Data validation checks included limiting responses within specific data ranges. While the majority of validation occurred on entry, some historically submitted erroneous data (eg, year 2102) were excluded. An update to the Registry platform (underway in November 2024) means that it will no longer be possible to enter erroneous data which falls outside strict validation limits.

Some data set amendments have been made over the last decade and fields added after the launch represent a smaller cohort (therefore the listed denominators vary). The recorded comorbidities are not exhaustive and the listed comorbidities are in online supplements 1 and 2.

Data analysis

Descriptive statistics were used to summarise data for this paper, focusing exclusively on mean, SD and summarised percentages. For the purpose of this analysis, increases and decreases over time were stated in absolutes, with no trend analysis conducted. Therefore, all analyses were conducted using Excel (MS Excel 365).

GAP staging was used as a marker of disease severity, named for and calculated using sex (gender, G), age (A) and lung function (physiology, P) details.[4]

RESULTS

Baseline characteristics

From January 2013 to February 2023, data were collected from 5052 cases. Of the 64 submitting hospitals (online supplement file 3), 13 are specialist centres in England directly commissioned by the National Health Service (NHS) to run a specialised ILD service. These are the only centres in England able to prescribe antifibrotic therapies for IPF. Of the remaining 51, 40 are secondary care centres in England and 11 are centres across the devolved nations (Scotland, Wales and Northern Ireland, where commissioning differs). The Registry is open to all respiratory physicians in the UK and participation was voluntary until it was mandated for specialist ILD centres in England in 2022. There were 3523 follow-up records representing 1516 unique patients, comprising 30% of all records. There was a median of two follow-up records per patient with a median of 523 days from presentation to follow-up.

The cohort had a mean of 22.2% females and 77.8% males (n=4886). The mean age at presentation was 74.0 (±8.1), with 73% (3524/4826) ≥70. Mean age increased from 71.0 to 75.7 in 2013–2022. Patients had a high burden of comorbidities at presentation, with 76% having ≥1 comorbidity. Hypertension and ischaemic heart disease

Table 1 Summarised UK IPF Registry data from January 2013 to February 2023. These figures are correct for the data cut taken in February 2023. Data may be entered retrospectively as well as prospectively, therefore future analyses may have slight variations in their totals

	N (total=5052)	Mean	±SD
Demographics			
Age in years	4826	74.0	8.1
Male (%)	4886	77.8	–
First-degree relatives with IPF (%)	4478	5.9	–
Arterial hypertension (%)	3857	39.9	–
Wait from referral to first clinic visit (weeks)	4480	13.6	15.0
Baseline physiology			
FEV1% pred.	4363	80.9	20.8
FVC% pred.	4370	77.0	24.1
D_{LCO}% pred.	3078	53.7	24.8
KCO% pred.	2926	80.7	27.9
GAP stage I (%)	3133	38.3	–
GAP stage II (%)	3133	52.4	–
GAP stage III (%)	3133	9.3	–

First-degree relative=parents, full siblings or children.
D_{LCO}, diffusion (transfer) factor for carbon monoxide; FEV1, forced expiratory volume in 1 s; FVC, forced vital capacity; GAP, a marker of severity calculated using sex (G), age (A) and physiology (P) data; IPF, idiopathic pulmonary fibrosis; KCO, carbon monoxide transfer coefficient.

accounted for 39.9% and 24.1% of cases respectively, followed by diabetes (22.3%) and hiatus hernia (20.8%). 66% were ex-smokers (2829/4281). 93.9% had no relatives with IPF (3017/3213), while 5.9% (188) had at least one first-degree and 0.3% (8) had at least one second-degree relative. First-degree relatives include parents, siblings and children. The second degree includes other biological relations, such as grandparents, aunts/uncles or nieces/nephews.

Mean forced vital capacity (FVC) was 77% predicted (±24.1) and diffusing capacity (DLCO) 54% predicted (±24.8) at presentation. 37% of patients had an FVC≥80% predicted, although patients with FVC≥80% displayed a mean reduced DLCO of 58% predicted. At presentation, the overwhelming majority of patients had GAP Stage I (38%, 1201/3133) or II (52%, 1642/3133) disease. 10% (219/2317) had previously been confirmed to have pulmonary hypertension or right heart strain, secondary to their lung disease, confirmed on echocardiogram or right heart catheterisation. The UK IPF Registry data relating to baseline characteristics are available in table 1.

Referral and wait times

60% (2561/4265) of patients reported chest symptoms for>12 months before the first clinic visit. 23% (996/4265) of all patients reported symptoms from 12 to 24 months before the first clinic visit and 37% (1565/4265) for>24 months. Referrals from secondary care respiratory physicians rose sharply from 50% to 69% in 2013–2014 (figure 1). This increase continued gradually, alongside decreased referrals from primary care (40–15% in 2013–2022).

The mean wait from referral to first clinic visit was 13.6 weeks; 13.9 for ILD specialist centres in England and 12.8 for all other centres. For specialist centres, waits were reduced by approximately 3 weeks from 2013 to 2015, before increasing by approximately 12 weeks by 2019. For all other non-specialist centres, waits were reduced by approximately 2 weeks from 2013 to 2016, before increasing by 12 weeks by 2019. From 2019–2022 waits at all centres gradually reduced by 5–6 weeks (figure 2).

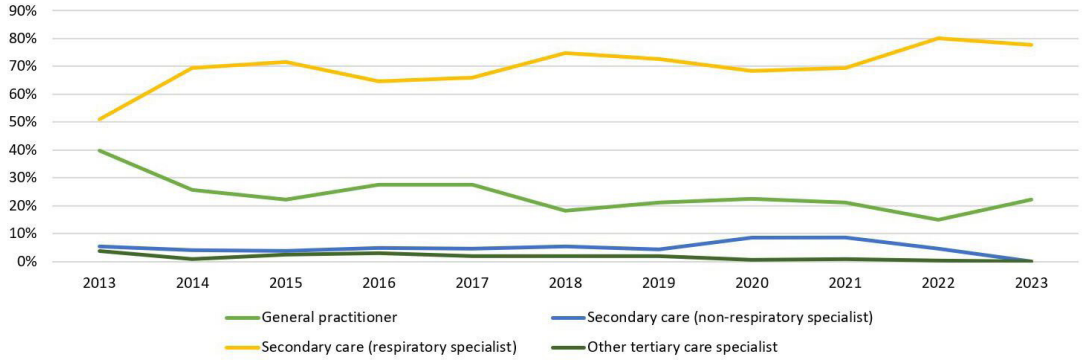
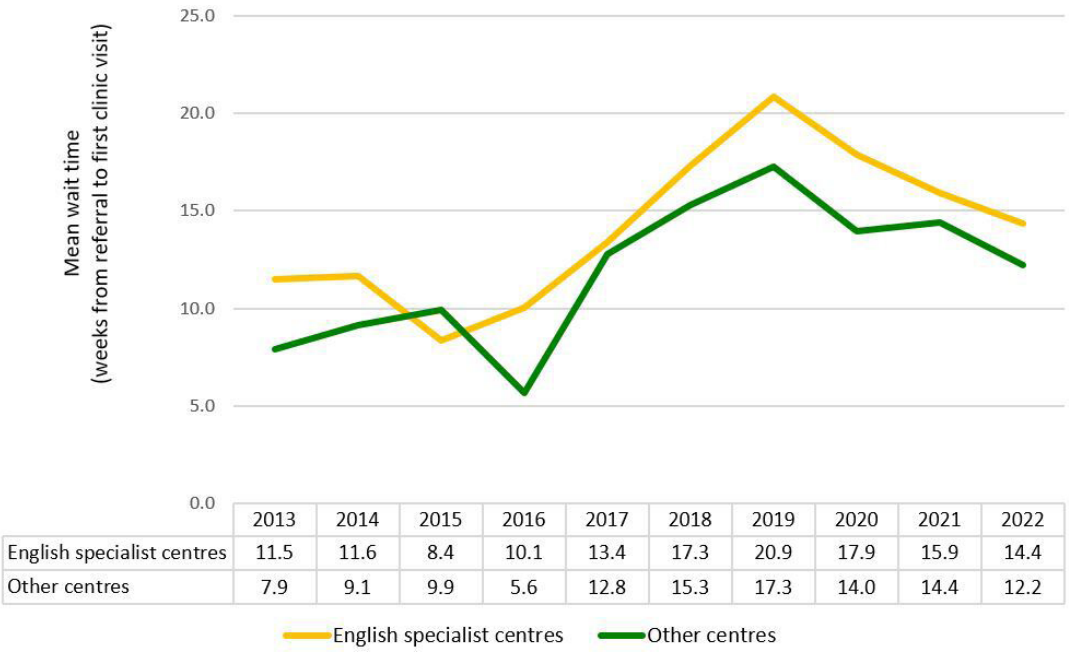


Figure 1 Route of referral to clinic over time. Since 2013, the primary route of referral has been directly from a respiratory specialist in secondary care. There appears to have been an increase in referral from respiratory specialists in secondary care (from 50.5% in 2013 to 78.4% in 2022) and a comparable reduction in referral from general practice (from 39.4% in 2013 to 14.5% in 2023). This may be due to the introduction of antifibrotic therapy—for which referral to tertiary care is required—and may have had an impact on waiting times.



	2013	2014	2015	2016	2017	2018	2019	2020	2021	2022
English specialist centres	11.5	11.6	8.4	10.1	13.4	17.3	20.9	17.9	15.9	14.4
Other centres	7.9	9.1	9.9	5.6	12.8	15.3	17.3	14.0	14.4	12.2

Figure 2 Wait times from referral to first clinic visit over time. This figure shows mean wait times (in weeks) from the date on the referral letter to the date of first clinic visit, by year of first clinic visit. English specialist centres have longer wait times than all other centres (ie, English non-specialist centres and centres from the devolved nations), though all centres share a similar pattern of increases/decreases in wait times over time.

Diagnosis

Patients were diagnosed using high-resolution CT, with 97% reporting definite or probable usual interstitial pneumonia (UIP) patterns. Overall, 5.6% (282/5052) required video-assisted thoracoscopic biopsy to confirm diagnosis, decreasing from 10.4% in 2013 to 3.9% in 2022.

Most cases were discussed at the ILD multidisciplinary team (MDT) meeting, with 93% discussed before the first clinic visit and 2% due for discussion at an upcoming MDT. Of the 93% of cases confirmed by MDT, 45% were diagnosed as having a definite, 30% likely and 18% as working diagnosis of IPF.

Management and outcomes

Figure 3 shows the proportion of patients prescribed drug treatments. The use of proton pump inhibitors remained consistent (35%), as did the proportion not receiving/prescribed drug treatment at presentation (33%). Figure 4 shows the proportion receiving pirfenidone or nintedanib since 2016. Over the decade 48% of patients were prescribed antifibrotic treatment at presentation and from 2017 nintedanib use superseded pirfenidone (in 2022 32% received nintedanib and 25% pirfenidone).

At presentation, 90% (1744/1937) had oxygen needs assessed, with 60% (1047/1744) not currently requiring oxygen therapy. 16% (647/4113) were receiving/prescribed at least one form of oxygen therapy at presentation, with 15% on ambulatory, 6% long-term and 1% short burst oxygen. Clinical trial recruitment at presentation remained consistently low at 7.5% (272/3603). The proportion deemed ineligible for lung transplantation 'at any time' at presentation (ie, the clinician has not only determined that they are ineligible for transplantation but that their eligibility status will not change) increased from 61% to 74% in 2013–2022. The overall mortality rate is 31%.

The National Institute of Clinical Excellence (NICE) Quality Standard for IPF[5] includes five quality statements. Figure 5 summarises data against each statement. 93% were discussed at MDT at presentation and a further 2% were due for discussion at an upcoming MDT. At presentation, 89% (1967/2211) were offered interaction with an ILD specialist nurse. 85% (1347/1580) of follow-ups assessed oxygen needs, of which 63% did not currently require oxygen therapy (847). At presentation, 89% (2494/2792) had pulmonary rehabilitation (PR) needs assessed, with 57% subsequently referred for PR. Reasons for not referring included the patient declining or having completed a course of PR <12 months ago (7% and 4% respectively), while 28% (687) were not suitable for referral, due to poor mobility or already having a good fitness level. At presentation, 82% (2444/2993) had palliative care needs assessed.

DISCUSSION

Patient registries are invaluable in providing 'real life' information on relatively uncommon conditions over time. The initial publication of this cohort covered 2013–2019, where demographic, diagnostic and treatment data were reported for 2474 patients.[3 6]

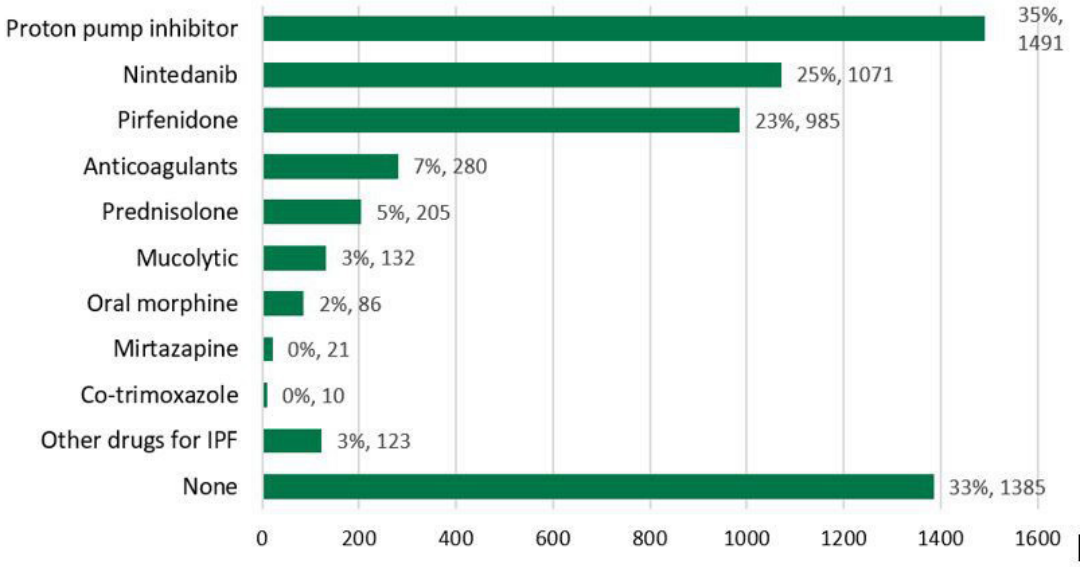


Figure 3 Drug treatments at presentation (including those prescribed at first clinic visit). This figure shows the drug treatments prescribed to patients at presentation. Pirfenidone has been available through the NHS since 2013, whereas nintedanib has only been available since 2016. The drug treatment options were expanded in December 2019. The option of 'warfarin' was changed to 'anticoagulants' in December 2019 which means the apparent increase in the use of anticoagulants may simply reflect the inclusion of other anticoagulant agents in this question. IPF, idiopathic pulmonary fibrosis; NHS, National Health Service.

This population resembles other cohorts. The male preponderance (78%) reflects a range of cohorts, although the mean age (74) is approximately 4–5 years older.[7] Registry data are largely in line with other publications, including epidemiological data on cardiovascular comorbidities.[8] The cohort had a high burden of comorbidities (76%≥1 comorbidity), with arterial hypertension (40%) broadly in keeping with the eurIPFreg (European IPF Registry and Biobank, 32%) and the German INSIGHTS-IPF Registry (54%).[9 10] The proportion with ischaemic heart disease (24%) closely resembled the cohorts of the IPF-PRO (Idiopathic Pulmonary Fibrosis Prospective Outcomes), INSIGHTS-IPF and eurIPFreg registries (18%, 25% and 29%).[9–11] Additionally, the proportion with at least one relative with IPF was 6%, while other cohorts vary widely, typically ranging from 4% to 25%.[12–17]

One concern with Registries worldwide is the certainty of diagnosis. The Latin American Registry of Idiopathic Pulmonary Fibrosis, which evaluated 761 patients from 14 countries,[18] used a three-tier verification system to ensure that the data uploaded did meet the diagnostic

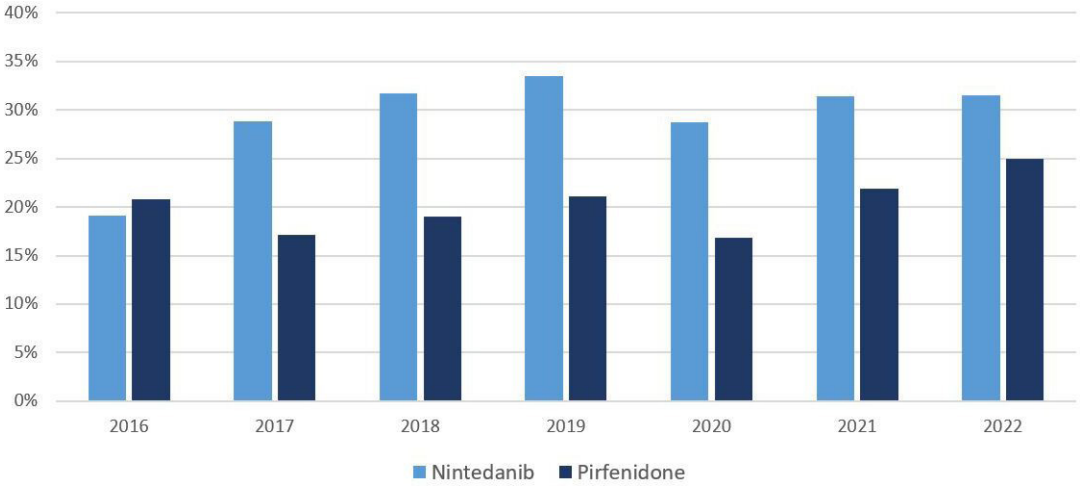


Figure 4 Proportion of patients receiving antifibrotic therapies at presentation. This chart shows a direct comparison of the proportions of patients receiving nintedanib and receiving pirfenidone at presentation since 2016, when nintedanib use was approved by NICE. Some patients were receiving both nintedanib and pirfenidone in the 3 months up to the first clinic visit (including drugs newly prescribed at the first clinic visit). NICE, National Institute of Clinical Excellence.

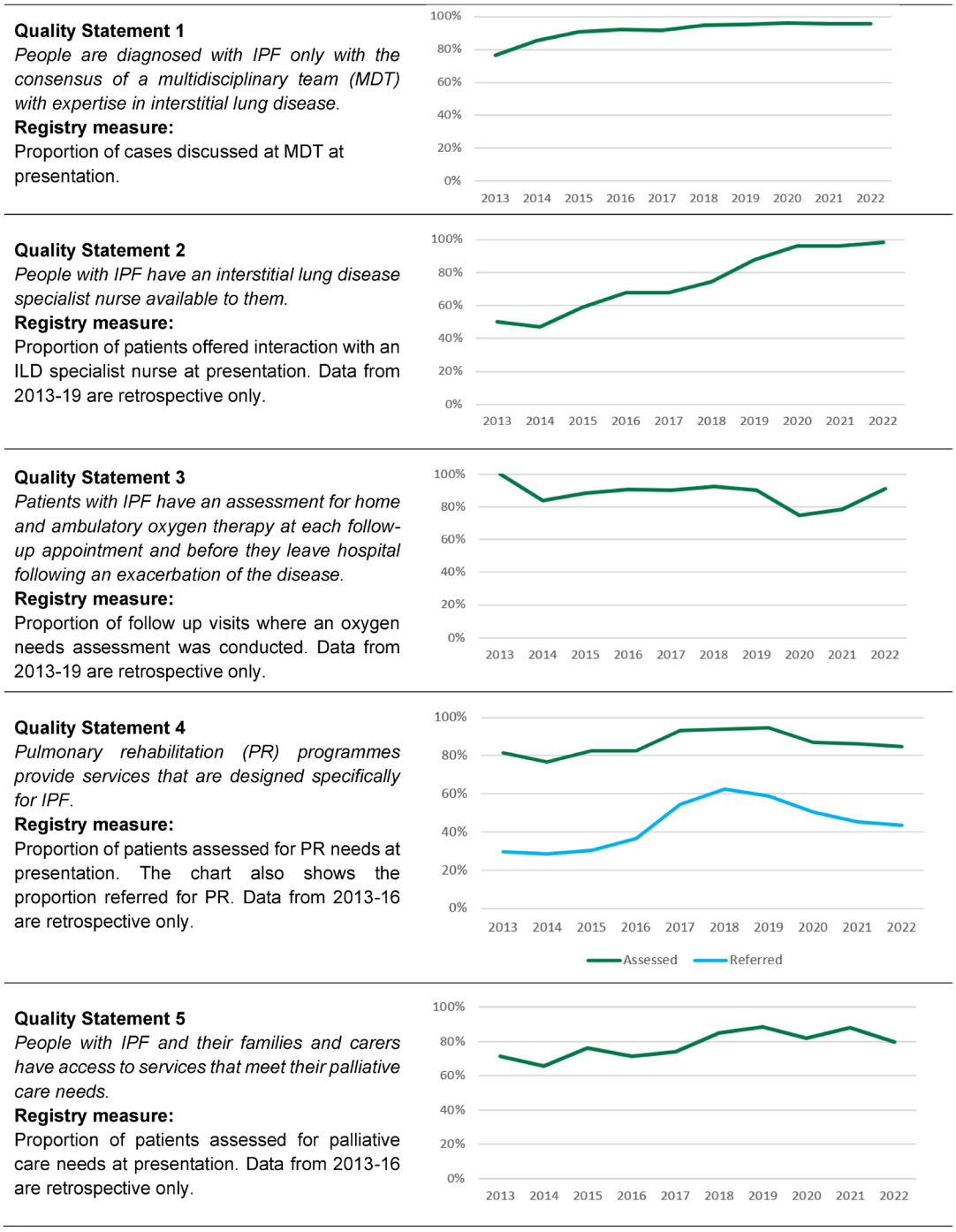


Figure 5 Performance against the five statements in the NICE Quality Standard for IPF. ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; NICE, National Institute of Clinical Excellence.

criteria for IPF according to the international guidelines produced collaboratively bythe American Thoracic Society, the European Respiratory Society, the Latin American Thoracic Association and the Japanese Respiratory Society (ATS/ERS/ALAT/JRS).[19] It is commendable how the system of verification was developed in this study, with a central expert committee reviewing all cases and ultimately excluding 213 from the initial submission of 974. For the UK IPF Registry, it was a prerequisite to enter data only if the diagnosis of IPF was made according to the internationally accepted ATS/ERS criteria and the majority of cases were confirmed to have IPF following specialist MDT discussion. That the cohort presented by the UK IPF Registry closely aligns with the characteristics of the Latin American cohort (which had, for example, 74.7% males, mean age of 72) is encouraging.

Patients experience lengthy waits from symptom onset to the first clinic visit. 23% had symptoms for 12–24 months and 37% for>24 months. This is longer than IPF-PRO, where only 49% were diagnosed >12 months after

symptom onset,[12] while a study in the USA reported an average time to diagnosis of 2.7 years after initial respiratory diagnosis.[20] The lengthy wait to diagnosis, coupled with the mean wait to be seen in the clinic of 13.6 weeks, highlights the increasing challenges to timely assessment and diagnosis. ILD services across the UK are challenged with inadequate workforce, directly impacting waiting times for outpatient assessment in ILD clinics. Waiting time from initial referral is a significant issue throughout the NHS, particularly in IPF, where patients are typically assessed in secondary care and wait again for a tertiary referral. While barriers to timely diagnosis are well documented,[21] long diagnostic waits are particularly distressing in a condition with high symptom burden and poor median survival.[22] UK IPF Registry data show even delays of 6–12 months result in significant survival disadvantage.[23] Other data sets also show time to referral associated with worse survival.[24 25] The strong links between delayed diagnosis and its deleterious effect on survival indicate overcoming challenges faced by the national workforce[26] is an essential component of improving care.

The rise in referrals from secondary care specialists from 2013 to 2014 (51–69%) may be a direct result of pirfenidone approval by NICE in 2013. It is unclear why the proportion of primary care referrals has been decreasing over time (40–15% in 2013–2022) and these data should serve as justification to evaluate this in detail at the local and national levels. However, it should be noted that specialist centres in England submitted 77% of records and the primary referral base from ILD specialist centres is from secondary care (rather than primary care). This highlights the limitations of having a voluntary, unfunded national registry. The low proportion of general practice referrals may negatively impact wait times in a way which is not reflected here, as waits following referral from secondary care do not take into account initial waits for referral from primary to secondary care.

Baseline characteristics were broadly in line with published data, with a mean FVC of 77% predicted compared with 68–81% for a number of cohorts.[7] The mean DLCO of 54% predicted was slightly higher compared with 36–50% for a number of cohorts.[7] GAP staging proportions (Stage I, 38%; Stage II, 52%; and Stage III, 9%—totalling 99% due to rounding error) represented less advanced disease than the INSIGHTS-IPF Registry (20%, 50% and 30%).[10] This is likely a result of treatment restrictions within the UK. In England, Wales and Northern Ireland, patients with FVC <50% predicted were less likely to be referred 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% have an FVC <50% predicted. Similarly, it is likely that the under-representation of GAP stage III (9%) is due to selection bias, as 77% of cases are from specialist centres in England, where severe cases are unlikely to be referred. As registry uptake among non-specialist centres in England increases, it is anticipated selection bias will be reduced.

Over the first decade of the registry, changes in practice over time became evident. The proportion of cases involving biopsy decreasing from 10.4% to 3.9% meant there was a high diagnostic rate on radiological grounds without the need for tissue for a confident diagnosis. This is likely a direct result of the updated clinical practice guideline published in 2018[19] which states biopsy is not required where probable/definite UIP is identified. The COVID-19 pandemic would also likely be a contributing factor since 2020. The proportion receiving at least one antifibrotic at presentation increased from 36% to 56% in 2013–2022. The proportion receiving antifibrotic therapy at presentation varies significantly by country, between 26% and 71%.[15 27–30] However, some enrolment windows included therapies started up to 6 months after diagnosis, whereas the UK figure includes cases up to the time of presentation only.

Until 2023, suitability for antifibrotic treatment was partly based on criteria defined by NICE (access in England and Wales limited to FVC 50–80% predicted). Nintedanib use superseded pirfenidone by a small proportion, despite later approval by NICE. Other factors, such as renal or liver function abnormalities, can preclude the use of antifibrotics. In December 2019, clinicians were asked why antifibrotic therapy was not prescribed at the presentation. For 53% of patients, this was because the FVC was outside 50–80% predicted, while 14% took time to consider their options. A small number of centres were unable to prescribe antifibrotic treatment. As 37% had FVC >80% predicted at presentation, this was a major factor in non-prescription. Nintedanib has been made available for these patients since May 2023[31] and we expect to see a marked increase in prescriptions.

As antifibrotic prescription is restricted to specialist ILD centres in England, there is a significant delay in patients accessing specialist ILD clinics to commence one of the two disease-modifying drugs proven to stabilise lung function in IPF. However, there are regional variations to this approach and shared care agreements between English specialist centres and associated District General Hospitals exist, where diagnosis is established following a regional MDT and patient care remains within the referring hospitals.

Despite changing practices over time, two areas of management remain low; lung transplantation and clinical trial recruitment. A significant proportion were deemed ineligible for transplantation (rising from 61% to 74% in 2013–2022). This could be partly due to the advanced age of the cohort (mean 74). From the cohort only 12 patients were confirmed to have received a transplant; however, as follow-up data are incomplete (31% of cases included ≥1 follow-up) this is unlikely to be reflective of the full population. Similar cohorts reported the proportion of patients assessed and listed for transplantation remains small.[9 32 33] Clinical trial recruitment has

been consistently low (7.5%), reflecting challenges faced by the ILD community when confronted with a disease having limited therapeutic options and no cure. Comorbidities also contribute to trial eligibility and future IPF trials should focus on adaptive multi-intervention platforms to improve recruitment and patient care.[34]

As a real-world database started a decade ago, there are several limitations. One is the lack of robustness in survival data, as participating centres are not always informed of a patient's death. Another is limited follow-up records, available for only 31% of patients. Patients are predominantly (77%) entered from specialist prescribing centres in England, so the population may be skewed towards a different patient group to those seen exclusively in secondary care. Voluntary data entry is also dependent on local resource and the COVID-19 pandemic has had a significant impact on registry input in 2020–2021. In 2022, the National Health Service England mandated the Registry for IPF cases at specialist ILD centres in England and since 28% of cases were entered during the last 14 months, this seems to have had an impact on uptake.

In conclusion, registries provide pertinent data for uncommon conditions, offering a means of understanding clinical and demographic characteristics in real-world scenarios, as opposed to randomised clinical trials where patients may be excluded due to pulmonary function, age or other characteristics. Delays in diagnosis, lack of access to treatment and lack of involvement in research are all known to have a negative impact on care.[35] It is paramount to have large-scale, longitudinal data sets for patients with ILDs such as IPF because without such data it would be challenging to identify and quantify unmet needs. Furthermore, registries can be useful tools to support business cases for services to support funding at local and national levels, with a view to improving the quality of life for patients with this incurable disease and high unmet needs.

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Acknowledgements The British Thoracic Society would like to acknowledge the contribution of staff at all 64 sites which submitted data (these sites are listed in Supplement 3) and the patients who gave consent for their anonymised data to be used.

Contributors All authors discussed and contributed to this paper which was prepared on behalf of the British Thoracic Society. AF and ML were responsible for the initial drafting of the manuscript and AF is the guarantor. ML was responsible for data analysis. IS, NC and AMW provided additional insight and revisions. ML is the Lung Disease Registry Programme Manager and is the member of staff at the British Thoracic Society responsible for the management of the UK Interstitial Lung Disease (ILD) Registry study and its online platform. AF, IS, SA, HA, LC, NC, SF, SH, L-PH, CH, PM, EP and AMW were all members of the UK ILD Registry Steering Group, the body responsible for advising on study planning, content and design and all had input into planning and preparing this paper. All authors reviewed and approved the revised manuscript being submitted.

Funding The British Thoracic Society wholly funds the UK Interstitial Lung Disease Registry. The Healthcare Quality Improvement Partnership (HQIP) provided a £20 000 starter grant (2011–2013). Boehringer Ingelheim and InterMune each provided a £6000 grant (£12 000 overall) to support the development of additional electronic features in 2014.

Competing interests None declared.

Patient and public involvement Patients and/or the public were involved in the design, or conduct, or reporting, or dissemination plans of this research. Refer to the Methods section for further details.

Patient consent for publication Not applicable.

Ethics approval This study is a non-interventional database study. It was approved by the East of England—Cambridge East REC (reference 22/EE/0235). Participants gave informed consent to participate in the study before taking part.

Provenance and peer review Not commissioned; externally peer reviewed.

Data availability statement Data may be obtained from a third party and are not publicly available. Researchers may apply to access UK Interstitial Lung Disease Registry data directly from the British Thoracic Society through the Society's data access request process. For further information please visit https://www.brit-thoracic.org.uk/quality-improvement/bts-clinical-data-policy-and-data-access/.

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