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Original Research

Assessment of the impact of social deprivation, distance to hospital and time to diagnosis on survival in idiopathic pulmonary fibrosis

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ABSTRACT

Background: Idiopathic pulmonary fibrosis (IPF) is a progressive condition associated with a variable prognosis. The relationship between socioeconomic status or distance travelled to respiratory clinics and prognosis is unclear.

Research question: To determine whether socioeconomic status, distance to hospital and time to referral affects survival in patients with IPF.

Study design and methods: In this retrospective cohort study, we used data collected from the British Thoracic Society Interstitial Lung Diseases Registry, between 2013 and 2021 (n = 2359) and calculated the quintile of Index of Multiple Deprivation 2019 score, time from initial symptoms to hospital attendance and distance as the linear distance between hospital and home post codes. Survival was assessed using Cox proportional hazards models.

Results: There was a significant association between increasing quintile of deprivation and duration of symptoms prior to hospital presentation, Gender Age Physiology (GAP) index and receipt of supplemental oxygen and antifibrotic therapies at presentation. The most deprived patients had worse overall survival compared to least deprived after adjusting for smoking status, GAP index, distance to hospital and time to referral (HR = 1.39 [1.11, 1.73]; p = 0.003). Patients living furthest from a respiratory clinic also had worse survival compared to those living closest (HR = 1.29 [1.01, 1.64]; p = 0.041).

Interpretation: The most deprived patients with IPF have more severe disease at presentation and worse outcomes. Living far from hospital was also associated with poor outcomes. This suggests inequalities in access to healthcare and requires consideration in delivering effective and equitable care to patients with IPF.

1. Introduction

Idiopathic pulmonary fibrosis (IPF), a chronic progressive scarring lung disease, has a worldwide prevalence of between 0.3 and 4.5 per 10,000, and it is estimated that 32,500 people live with IPF in the UK [1,2]. Treatment options are limited but include antifibrotic therapy, oxygen and pulmonary rehabilitation [3–5] and survival remains poor with a median survival of 5.7 years [6]. This is variable with prognosis related to numerous factors including age, gender, lung function (captured in the Gender, Age and Physiology [GAP] index), hospitalisation, and pulmonary hypertension [7–9].

Socioeconomic status (SES) is an important independent determinant of health outcomes. It is defined as an individual's or group's social or economic standing, or positioning on the socioeconomic scale, and comprises characteristics including income, education, occupation and place of residence [10]. The relationship of disadvantaged SES to poor health outcomes in respiratory disease is recognised [11–13]. Likewise, the distance patients travel to attend hospital has a negative influence on

Abbreviations: BTS, British Thoracic Society; FVC, forced vital capacity; GAP, Gender Age Physiology; HR, Hazard ratio; IMD, Index of Multiple Deprivation; ILD, Interstitial Lung Disease; IPF, Idiopathic Pulmonary Fibrosis; IPF-PRO, Idiopathic Pulmonary Fibrosis Prospective Outcomes; SES, Socioeconomic status.

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health outcomes. A cohort study (n = 129) demonstrated that people with chronic obstructive pulmonary disease living in isolated rural areas have worse survival than those living in urban areas [14].

There are limited robust data on the effect of SES on interstitial lung disease (ILD) outcomes and care, especially in universal healthcare systems. The most disadvantaged people with progressive pulmonary fibrosis living in the USA, but not Canada, have a higher mortality rate with a hazard ratio of 1.51, with similar findings in the IPF cohort (n = 1606) [15]. This is in contrast to data from the multi-centre US Idiopathic Pulmonary Fibrosis Prospective Outcomes (IPF-PRO) Registry which did not show that income was associated with death, but higher income was related to the chance of receiving a lung transplant, in 955 patients [16]. Rural patients with IPF, from the University of California, San Francisco ILD Cohort (n = 843), were more breathless, had more oxygen use and lower lung function when presenting to ILD speciality care compared to urban patients but there was no significant difference in survival between rural and urban patients [17]. The French COFI Registry, which captured city level scoring of SES, reported that, although there was no difference in survival, those with lower incomes (n = 50) had worse progression free survival (11.6 vs. 16.0 months) than those with higher incomes (n = 150) [18]. In terms of travelling distance, the IPF-PRO Registry reported this had a weak association to death over a 2-year period [16]. Patients with fibrotic ILD living more than 70 km away from a specialist centre had 50 % increase in risk of having a transplant or dying (hazard ratio [HR]: 1.52; 95 % confidence interval [CI], 1.10–2.11) [19]. These studies do not examine process, have small sample sizes, are single centre studies or do not adjust adequately for potential confounders.

The British Thoracic Society (BTS) ILD Registry is an ethically approved disease specific registry capturing data from people with IPF (and sarcoidosis) regarding demographics, referral patterns, treatment, and outcomes [20]. IPF is confirmed following local multidisciplinary team meeting according to contemporaneous international guidelines [21,22]. Contribution is voluntary but participants are required to provide written informed consent prior to doing so. Although the registry is open to all hospitals in the UK, only the specialist centres in the UK are expected to contribute patients and as such much of the data reflects tertiary care. The aim of our study was to determine whether SES and distance patients live from their specialist hospital affects survival outcomes (Overall Survival and Progression Free Survival) in patients with IPF using data from the BTS ILD registry. Secondary aims are to estimate the relationship between SES and distance with baseline demographics, lung function, time to referral and provision of IPF care.

2. Methods

2.1. Ethics

This is a retrospective cohort study of people with IPF in England. Data were obtained from the BTS ILD Registry, following review and approval by the BTS Information Governance Committee, and data sharing agreements on July 9, 2021 (reference 007/21). The dataset collected for the BTSILD Registry programme was granted ethical approval for epidemiological research by the NRES Committee East of England in 2012 (17/EE/0346) and this was renewed in October 2017 [23]. The University of East Anglia Faculty of Medicine and Health Sciences Research Ethics Committee (UEA FMH REC) also approved this study (Application ID: ETH2122-0797).

2.2. Patients

All patients were diagnosed with IPF, following multidisciplinary team meeting, according to contemporaneous international guidelines. Patients were confined to those living in England due to the small number of individuals from other devolved nations and the differences in the definition of SES throughout the UK. Patients with missing demographic, baseline lung function data and those whose Index of Multiple Deprivation (IMD) score and distance travelled to respiratory clinic could not be defined were excluded.

2.3. Dataset

Data were available from February 2013 until June 2021. Duration of symptoms prior to first review in respiratory clinic was recorded in a categorical manner: no symptoms, less than 6 months, 6–12 months, 12–24 months or more than 24 months. Provision of oxygen and pulmonary rehabilitation are recorded as “referred or receiving”, “patient declined”, “patient not suitable”, “not required” or “not assessed” in the BTS ILD Registry. Reasons for not starting antifibrotic therapy were scored as “not meeting prescribing criteria”, “patient choice”, “renal impairment”, “liver impairment”, “decision deferred by patient”, or “other” in the BTS ILD Registry. For the purposes of the analysis, “patient-related reasons” for not starting antifibrotic therapy were taken as either of “patient choice” or “patient wants to consider options” responses from the BTS ILD Registry and “clinical-related reasons” were taken as the sum of “not meeting prescribing criteria”, “renal impairment” and “liver impairment”. Time to diagnosis was calculated as the date of onset of symptoms (above) to date of multidisciplinary team meeting. Lung function was obtained from hospital lung function laboratories at the time of first review at the respiratory clinic with percent predicted obtained from local algorithms. Data were stored in password protected files on secure servers within Norfolk and Norwich University Hospital and University of East Anglia. Access to the data was limited to the research team.

2.4. Outcomes

The primary outcome was overall survival defined as time from date of diagnosis to death from any cause. Clinical service-related outcomes include time to diagnosis and provision of healthcare resources (oxygen therapy, pulmonary rehabilitation and anti-fibrotic therapy). Patient-related outcomes included demographic characteristics such as age, sex, smoking status, comorbidities and uptake of therapies.

2.5. Exposures

The primary exposures of interest in our study was social deprivation as measured by the Index of Multiple Deprivation (IMD) 2019), time to referral and distance from home to respiratory clinic. The IMD 2019 (https://www.gov.uk/government/statistics/english-indices-of-deprivation-2019) encompasses seven main types of deprivation: “income, employment, education, health, crime, access to housing and services, and living environment” and is determined at neighbourhood level. Patients were categorised into 5 groups based on UK population level quintiles, as population-based quintiles are available and permits external comparison. Time was defined as duration of symptoms prior to attendance at clinic and was treated as a categorial variable: no symptoms, less than 6 months, 6–12 months, 12–24 months or more than 24 months. The distance from home to hospital was calculated as the linear distance between the respective postcodes using this tool: https://www.freemaptools.com/distance-between-uk-postcodes.htm.

2.6. Statistical analysis

A comparison of demographic and clinical characteristics was made across the deprivation quintiles. A test-for-trend across deprivation quintiles was performed using Pearson’s correlation between age, percent predicted forced vital capacity (FVC), percent predicted diffusing coefficient for carbon monoxide (DLCO) and the deprivation quintiles. For comorbidities and other binary variables, the chi-squared test for trend was conducted. The association between deprivation quintiles and distance quintiles at baseline was based on a linear-by-

linear test.

The association between the baseline characteristics and time-to-death was undertaken using Cox-Proportional Hazards regression models, including a random-effect term by hospital to account for potential different survival rates between hospitals. As standard for survival analysis model a gamma distribution was assumed for the random effect. The reference levels were those least deprived, shortest time to referral and closest to hospital. Individuals were followed up until death, however individuals were censored at the last lung function test. First, an unadjusted association was estimated for each factor. Then to remove potential confounding an adjusted analysis (adjusting for GAP score [7] classified according to GAP staging (0–3; 4 to 5; and 6 to 8), IMD quintile, distance quintile, time and smoking status) was estimated. In order to assess if there was an interaction between deprivation and distance an additional term for individuals most deprived and further away was included in a sensitivity model. The corresponding hazard ratios and 95 % confidence intervals are presented. The characteristics of patients who had versus those who did not have a follow-up data were compared using a linear test for trend for socioeconomic status quintile and time to referral and T-test for distance travelled. All significance tests were carried out at 5 % two-sided level of significance. Analysis was done using STATA 18.0.

3. Results

A total of 3398 patients from the BTS ILD Registry had sufficient baseline data available for inclusion in the analyses. However, patients from Scotland (n = 50), Wales (n = 27) and Northern Ireland (n = 55) were excluded, along with those with an unrecorded nation (n = 9), because of small numbers and the differences in calculating deprivation between the devolved nations. Patients were also excluded if baseline forced vital capacity (missing n = 698) or demographics (n = 4) were missing or if IMD (n = 63), distance travelled to hospital (n = 18), or time to referral (n = 115) was not attainable/matched for a patient.

A total of 2359 patients were included in the baseline analysis (Table 1) with a mean (standard deviation) age of 74 [8] years and a male predominance (79.5 %). A significant percentage of patients from the most deprived quintile (IMD quintile 1) were either current smokers or ex-smokers (73 %) compared to patients who were in the least deprived quintile (66 %). Cardiovascular comorbidities were similar between the deprivation quintiles (Table 1).

There was a significant difference in the baseline lung function, with lower lung function in the most deprived IMD quintile. Those in the most deprived quintile had a higher GAP index (Table 1). In keeping with more severe disease in the most deprived quintile; a higher percentage in this group required supplemental oxygen therapy (Table 2). A higher proportion of patients in the most deprived quintile were referred to pulmonary rehabilitation (69 %) or were prescribed antifibrotic therapy (93 %) compared to the least deprived (46 % and 78 % respectively).

More patients in the most deprived IMD quintile presented more than two years after the onset of symptoms (42 %) compared to the least deprived quintile (36 %). Furthermore, fewer patients in the most deprived quintile were seen within six months. Those most deprived lived closer to the hospital than those least deprived (Table 1).

A total of 1380 patients had follow-up data available with a median length of follow up of 28 months (range 1–129 months). Of those 875 (63.4 %) died during follow-up. The median baseline values for those who had follow-up data versus those without follow-up data for

Table 1
Comparison of demographic and clinical characteristics of IPF patients by IMD quintile.

Characteristics[a]	No. (%)						
	Q1 (Most deprived)	Q2	Q3	Q4	Q5 (Least deprived)	Total	*P*
N	469	389	447	510	544	2359	
Age, mean (SD)	72 (8.57)	72.5 (8.12)	73.7 (7.63)	74.7 (7.82)	74.8 (7.66)	73.6 (8.03)	**<0.0001**[c]
Sex, Male	363 (77.4 %)	299 (76.9 %)	357 (79.9 %)	416 (81.6 %)	440 (80.9 %)	1875 (79.5 %)	**0.047**[b]
Smoking status							
Current smoker/ex-smoker	312 (72.6 %)	267 (73.4 %)	287 (67.7 %)	333 (68.7 %)	342 (65.6 %)	1541 (69.3 %)	**<0.0001**[b]
Never/negligible	118 (27.4 %)	97 (26.6 %)	137 (32.3 %)	152 (31.3 %)	179 (34.4 %)	683 (30.7 %)	
Comorbidities							
IHD	115 (24.5 %)	69 (17.7 %)	101 (22.6 %)	109 (21.4 %)	109 (20.0 %)	503 (21.3 %)	**0.146**
GORD	77 (16.4 %)	73 (18.8 %)	86 (19.2 %)	116 (22.7 %)	115 (21.1 %)	467 (19.8 %)	**0.019**
DM	109 (23.2 %)	78 (20.1 %)	82 (18.3 %)	107 (21.0 %)	100 (18.4 %)	476 (20.2 %)	**0.173**
Hypertension	164 (35.0 %)	125 (32.1 %)	162 (36.2 %)	189 (37.1 %)	190 (34.9 %)	830 (35.2 %)	**0.538**
GAP index mean (SD)							
	4.66 (1.48)	4.49 (1.39)	4.44 (1.49)	4.42 (1.47)	4.27 (1.43)	4.45 (1.46)	**<0.0001**[c]
GAP stage							**<0.001**[c]
0–3	99 (21.1 %)	97 (24.9 %)	129 (28.9 %)	157 (30.8 %)	188 (34.6 %)	670 (28.4 %)	
4–5	227 (48.4 %)	198 (50.9 %)	211 (47.2 %)	225 (44.1 %)	239 (43.9 %)	1100 (46.53 %)	
6–8	143 (30.5 %)	94 (24.2 %)	107 (23.9 %)	128 (25.1 %)	117 (21.5 %)	589 (24.97 %)	
Lung function mean (SD)							
FVC	75.8 (17.8)	77 (17.4)	77.9 (17.3)	77.4 (15.9)	80.4 (17.9)	77.8 (17.3)	**0.0001**[c]
D_LCO	45.6 (14.5)	48.7 (14.7)	49.5 [14]	50.8 (14.9)	52.8 (15.8)	49.7 [15]	**<0.0001**[c]
Duration of symptoms prior to chest clinic							**0.015**[b]
<6 months	45 (9.7 %)	36 (9.4 %)	49 (11.3 %)	59 (11.9 %)	73 (13.8 %)	262 (11.4 %)	
6–12 months	107 (23.2 %)	93 (24.3 %)	105 (24.3 %)	104 (21.0 %)	132 (24.9 %)	541 (23.5 %)	
12–24 months	116 (25.1 %)	89 (23.2 %)	105 (24.3 %)	120 (24.2 %)	133 (25.1 %)	563 (24.4 %)	
>24 months	194 (42.0 %)	165 (43.1 %)	173 (40.0 %)	213 (42.9 %)	192 (36.2 %)	937 (40.7 %)	
Distance quintile							<0.001[b]
1 (Nearest) [≤4.0 miles]	169 (36.0 %)	90 (23.1 %)	64 (14.3 %)	83 (16.3 %)	67 (12.3 %)	473 (20.1 %)	
2 [>4.0–8.3 miles]	85 (18.1 %)	76 (19.5 %)	96 (21.5 %)	101 (19.8 %)	113 (20.8 %)	471 (20.0 %)	
3 [>8.3–14.0 miles]	81 (17.3 %)	55 (14.1 %)	85 (19.0 %)	114 (22.4 %)	137 (25.2 %)	472 (20.0 %)	
4 [>14.0–24.1 miles]	57 (12.2 %)	81 (20.8 %)	101 (22.6 %)	108 (21.2 %)	125 (23.0 %)	472 (20.0 %)	
5 (Furthest) [>24.1–85.1 miles]	77 (16.4 %)	87 (22.4 %)	101 (22.6 %)	104 (20.4 %)	102 (18.8 %)	471 (20.0 %)	

Abbreviations: DM, Diabetes Mellitus; GAP Index, Gender (G) Age (A) Pulmonary Physiological Parameter (P); GORD, Gastro-oesophageal reflux Disease; IHD, Ischaemic heart disease; Q1, quintile 1; Q2, quintile 2; Q3, quintile 3; Q4, quintile 4; Q5, quintile 5.

[a] All categorical variables are presented as No. (%).

[b] based on chi-squared test for trend.

[c] Based on Pearson correlation coefficient.

Table 2
Comparison of medical interventions and reasons for not receiving by IMD quintile.

Intervention	No. (%)					
	Q1 (Most deprived)	Q2	Q3	Q4	Q5 (Least deprived)	*P*
Oxygen						<0.001
Not assessed	5 (6.7 %)	2 (2.7 %)	3 (3.1 %)	6 (5.9 %)	8 (4.6 %)	
Referred/already receiving	35 (46.7 %)	17 (23.0 %)	24 (24.5 %)	20 (19.6 %)	21 (12.1 %)	
Patient declined	4 (5.3 %)	5 (6.8 %)	3 (3.1 %)	3 (2.9 %)	6 (3.4 %)	
Not required/appropriate	31 (41.3 %)	50 (67.6 %)	68 (69.4 %)	73 (71.6 %)	139 (79.9 %)	
Pulmonary rehabilitation						**<0.001**
Not assessed	8 (2.9 %)	14 (7.0 %)	21 (8.6 %)	15 (5.5 %)	24 (7.7 %)	
Assessed and referred	192 (69.1 %)	115 (57.2 %)	130 (53.3 %)	147 (53.8 %)	143 (45.8 %)	
Patient declined	63 (22.7 %)	60 (29.9 %)	76 (31.1 %)	98 (35.9 %)	130 (41.7 %)	
Not required/appropriate	15 (5.4 %)	12 (6.0 %)	17 (7.0 %)	13 (4.8 %)	15 (4.8 %)	
Antifibrotic therapy received						**<0.001**
Received within 3 months (of consultation)	438 (93.4 %)	356 (91.5 %)	392 (87.7 %)	445 (87.3 %)	426 (78.3 %)	
Antifibrotic therapy not given						**0.010**
FVC out of range/clinically inappropriate	169 (36.2 %)	147 (37.9 %)	185 (41.4 %)	209 (41.1 %)	253 (46.9 %)	
Other	8 (1.7 %)	9 (2.3 %)	13 (2.9 %)	8 (1.6 %)	20 (3.7 %)	
Patient declined	7 (1.5 %)	8 (2.1 %)	10 (2.2 %)	14 (2.8 %)	13 (2.4 %)	

socioeconomic status quintile, distance from hospital (miles) were: 3 [2–4] vs 3 [2–5], p < 0.001 and14.53 (12.7) vs 14.35 (13.09) p = 0.72. There was no association between time to referral and follow-up status.

Variables associated with worse survival included higher GAP index, increased time to referral, increasing deprivation, and longer distance travelled to hospital (Table 3). Patients from the most deprived quintile had a 36 % increase in the risk of death compared with those from the least deprived quintile, (HR = 1.36 [1.10–1.69]; p = 0.004). In the adjusted model (adjusted for age, smoking status, GAP index, time to referral and travel distance to hospital), the increased risk of death remained in the most deprived quintile (adjusted HR = 1.39 [1.11–1.73] p = 0.004). There was a significant difference in survival amongst the deprivation quintiles (p < 0.001).

Compared to patients living nearest to hospital those living further away had a 34 % increased risk of death (1.34 [1.06–1.69] p = 0.013; Table 3) in an unadjusted analysis. GAP index was independently associated with survival (Table 3).

Table 3
Estimated hazard ratios for overall survival by risk factor.

Factor	Unadjusted		Adjusted	
	HR (95 % CI)	*P*	HR (95 % CI)	*P*
Deprivation: IMD quintile				
1 (Most deprived)	1.36 (1.10,1.69)	0.004	1.39 (1.11,1.73)	0.003
2	1.11 (0.89,1.38)	0.376	1.05 (0.84,1.32)	0.666
3	1.13 (0.91,1.42)	0.271	1.1 (0.88,1.38)	0.411
4	1.14 (0.92,1.40)	0.229	1.06 (0.86,1.32)	0.585
5 (Least deprived)	1		1	
GAP Stage:				
0–3	1		1	
4–5	2.17 (1.79,2.63)	<0.001	2.17 (1.78,2.64)	<0.001
6–8	3.41 (2.74,4.23)	<0.001	3.42 (2.73,4.28)	<0.001
Delay:				
<6 months	1		1	
6–12 months	1.32 (1.01,1.72)	0.041	1.34 (1.02,1.77)	0.034
12–24 months	1.23 (0.94,1.61)	0.138	1.19 (0.9,1.57)	0.228
>24 months	1.27 (0.99,1.63)	0.065	1.21 (0.94,1.57)	0.139
Distance to hospital (miles):				
1 (Nearest) [≤4.0 miles]	1		1	
2 [>4.0–8.3 miles]	1.13 (0.90,1.43)	0.284	1.2 (0.95,1.52)	0.13
3 [>8.3–14.0 miles]	1.37 (1.09,1.71)	0.007	1.43 (1.13,1.8)	0.003
4 [>14.0–24.1 miles]	1.27 (1.01,1.59)	0.039	1.29 (1.02,1.64)	0.034
5 (Furthest) [>24.1–85.1 miles]	1.34 (1.06,1.69)	0.013	1.29 (1.01,1.64)	0.041
Smoking status:				
Current or ex-smoker	1.15 (0.99,1.35)	0.075	1.11 (0.95,1.3)	0.179
Never smoker	1		1	

4. Discussion

In this large national cohort of patients diagnosed with IPF we demonstrate that socioeconomic deprivation was independently associated with death in a universal healthcare system. Patients with increased social deprivation reported a greater time to assessment, and had more severe disease at presentation, with a higher GAP index compared to those who are least deprived. They were also more likely to have a smoking history. However, the association between outcomes and deprivation remained when adjusted for time to assessment and severity of disease (baseline GAP index) and is unlikely to be due to deficiencies in specialist treatment which was higher in this group. GAP index and distance from hospital were also associated with death when adjusted for the available confounding factors. Although longer time to referral was associated with higher risk of dying, this only reached significance for a duration up to 12 months. The likelihood of being followed-up was associated with higher socioeconomic quintile but not distance to hospital or time to referral, however the differences were small. Therefore, as social deprivation, travelling distance and time to referral all contribute to the increased disease severity at specialist centre review and overall poor outcomes, all of these elements need to be addressed.

The main strengths of this study are the size of the dataset and that all patients were diagnosed with IPF following expert multidisciplinary team discussion. This allowed adjustment for important confounders. To the best of our knowledge, this is the first study to assess the effect of SES, travelling distance and time to referral along with clinical-service provision and patient care at the same time in a universal health care system. However, this was a retrospective cohort study using clinically captured data. Data regarding survival were not confirmed from the Office of National Statistics and may be incomplete, however survival was similar to that expected for people with IPF and, as SES is unlikely to be related to data completeness, the findings for relative risk are reliable. The SES was measured using the Index of Multiple Deprivation as opposed to individual based scores, due to the lack of individual specific data. However, IMD has been proven to be the most reasonable proxy

[24]. Likewise, we used linear distance between the home and hospital postcode rather than road distance needed to travel or journey time, which may have more accurately captured patient burden, however linear distance is an acceptable surrogate for travelling distance or time [25,26]. Ethnicity was not assessed as part of the study but is recognised as a factor of poor outcome in many diseases however only 17 % of those recorded in the registry are of non-white race [23]. Likewise, we did not have access to data about lung transplant, however only about 1 % of individuals are referred for transplant in the BTS registry [23] with far fewer being accepted. Lead time bias may influence the results of our study as the time of entry of the data onto the registry was determined by the attending physician (those most deprived had more severe disease at entry and may have been diagnosed later) however, associations were independent of time to referral which is a measure of lead time bias. We did not convert all lung function values to percentage predicted values using Global Lung Index, rather we used locally derived normalised values as recorded in the registry, however we adjusted for site throughout the analysis to account for the potential difference in percent predicted derivations.

We showed that deprivation is important in a universal healthcare system, which is in keeping with a prospective study in France [18], but contradictory to the hypothesis from Goobie et al. [15] who showed deprivation related to mortality in USA but not in Canada and that of the IPF-PRO Registry, which showed that access to care (lung transplant) was related to socioeconomic status [16]. Our findings suggest that social deprivation exerts its effects on poor outcomes in addition to that of access to care and other aspects of social deprivation. In this respect, poor nutrition is associated with all-cause mortality [27], pollution is associated with decline in lung function [28] and Black and Hispanic people have higher age-adjusted mortality rate compared to White people [29] in IPF.

We showed that there was a non-linear relationship between deprivation and outcomes, with only the most deprived having a significant effect on outcomes. This is in keeping with Goobie et al. [15] who showed that the effect of social deprivation was mostly confined to the most deprived. Although a larger sample size may have identified significant differences between groups, other studies have shown more marked effects of deprivation in those most affected [30]. Similarly, we demonstrate that longer distance travelled to hospital is associated with worse all-cause mortality. Our findings are in keeping with those of Johannson et al. who found longer travelling distances were associated with poor outcomes in Canada [19]. Although rural living brings advantages to people with chronic respiratory illness, extensive travel and erratic physician availability are perceived as troublesome [31]. Greater uptake of home monitoring may reduce these difficulties but may widen the gap seen due to poor socioeconomic status [32].

We have shown that time from symptom onset to assessment in specialist clinics is common with 40 % of people waiting for at least two years. Others have reported high rates of respiratory symptom burden prior to diagnosis with nearly 80 % of patients having had a respiratory related primary consultation in the year prior to referral; advocating greater awareness of IPF in primary care [33,34]. Time to referral is also associated with worse survival in IPF [35–37]. In a small US study of 129 patients with IPF, Lamas et al. [35]. demonstrated worse survival is associated with time to specialist review and this was independent of insurance type and educational attainment. Male gender and initial misdiagnosis have been associated with increased time to specialist review [38]. In our study, we demonstrate that longer time to assessment is associated with increased deprivation and distance travelled to hospital and numerically higher but non-significant increase in deaths. In our study, the most deprived are also more likely to experience symptoms for longer before assessment in respiratory clinic.

Most of the patients in our study had a cigarette smoking history. Smoking has been shown to have a positive dose-response relationship with the risk of IPF and smoking is higher in disadvantaged groups [39,40]. It is possible that smokers may delay presentation with IPF as they do with lung cancer [41]. However, smoking history was not independently associated with adverse outcomes in our study and others have shown that it is not an independent risk factor for diagnostic delay in IPF [38].

We did not show that poor outcomes seen in the most deprived group were related to a longer time from symptom onset to review in a respiratory clinic; using different interaction adjustments (term for the most deprived and furthest away, a linear-by-linear interaction term or an additional linear term for the most deprived) resulted in hazard radios of between 0.98 and 1.01. However, those with lower socioeconomic status lived closer than those with higher socioeconomic status possibly masking the effect of travel. It is possible that patients from poor socioeconomic class, although living close to hospital, are only referred to specialist services when their lung function deteriorates, and specialist input is required thus the longer time to diagnosis identified in these patients is artefactual. However, our findings are in keeping with the finding of Ahuja et al. [42], who showed that socioeconomic status had a minimal effect on travel for people with a stroke. Limited access to healthcare resources may also play a role in the poor outcomes seen in the most deprived group [43].

The association we show between increase distance to respiratory clinics and reduced overall survival has important implications for the delivery of IPF care. The current hub and spoke model of care delivery in the NHS, with treatment provided by specialist centres, may result in impaired access to care. Care closer to home may result in greater survival. Our results also suggest that future screening programmes and policies need to be put into place to mitigate the differences in socioeconomic levels leading to poorer health outcomes in patients with IPF. Research is required to determine whether screening programmes, the potential provision of services for those more socioeconomically deprived and the ability to access care closer to home improves survival in IPF. Further assessments are also required to determine the reasons for delays to initial presentation at primary care and delays to referral and more detailed mapping of patient pathway and referral patterns in those living close to a spoke versus those living close to a hub. It is important to overcome all barriers to care given the progressive nature of IPF.

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Data reporting

Data access for the British Thoracic Society Interstitial Lung Disease Registry is available at https://www.brit-thoracic.org.uk/quality-improvement/bts-clinical-data-policy-and-data-access/

CRediT authorship contribution statement

Rashmi Shankar: Writing – original draft, Methodology, Investigation, Funding acquisition, Formal analysis. **Charaka M. Hadinnapola:** Writing – review & editing, Validation, Methodology, Investigation, Formal analysis. **Allan B. Clark:** Data curation, Formal analysis, Funding acquisition, Methodology, Supervision, Writing – review & editing. **Huzaifa Adamali:** Conceptualization, Investigation, Writing – review & editing. **Nazia Chaudhuri:** Conceptualization, Investigation, Writing – review & editing. **Lisa G. Spencer:** Conceptualization, Investigation, Writing – review & editing. **Andrew M. Wilson:** Conceptualization, Funding acquisition, Investigation, Methodology, Resources, Supervision, Writing – review & editing.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests:

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