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Baseline characteristics from the EXCITING-ILD registry

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ABSTRACT

Background Interstitial lung diseases (ILDs) comprise a group of more than 200 different subtypes. They vary widely in terms of incidence, prognosis and treatment, yet real-life data from Germany are sparse.

Methods The prospective Exploring Clinical and Epidemiological Characteristics of Interstitial Lung Diseases (EXCITING)-ILD registry included patients with all different ILD subtypes from different healthcare settings. Follow-up ranged from 36 months to 5 years. Data were analysed descriptively. Baseline characteristics, diagnostic and treatment information are presented as absolute numbers and percentages. The Wilcoxon signed-rank sum test was used to quantify differences between groups. Line plots and bar plots were used for graphical presentation.

Results A total of 601 patients (60.7% men, mean age 64.3 years) from 32 centres were included in the EXCITING-ILD registry. The most common subtypes were sarcoidosis with 26.6% (n=160) and idiopathic pulmonary fibrosis (IPF) with 25.3% (n=152). Pulmonary hypertension was present in 8.7% of patients (n=52), with high incidences in connective tissue disease-associated ILD (16.3%) and pneumoconiosis (27.3%). The mean forced vital capacity was 76.4% predicted, and the mean DLCO-SB (diffusing capacity for carbon monoxide) was 54.1% predicted. The mean time to diagnosis was 38.8 months (SD 64.4) and was significantly shorter when the diagnosis was made after multidisciplinary discussion (31.6 vs 49.2 months, p<0.001). The frequency of surgical lung biopsies decreased over time in the registry, whereas the proportion of cryobiopsies showed a notable increase. In IPF, the number of patients treated with antifibrotics increased from 35.2% before 2015 to 48.4% in 2019.

Conclusion The EXCITING-ILD registry describes the frequency of ILD subtypes, ILD-related impairments, selected comorbidities and diagnostic and treatment patterns in a representative German population.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Interstitial lung diseases (ILDs) are a group of more than 200 rare and very rare diseases. Incidence, mortality and disability-adjusted life years are increasing. Registry data including all ILD subtypes and taking into account the latest consensus classification are scarce.

WHAT THIS STUDY ADDS

⇒ The Exploring Clinical and Epidemiological Characteristics of Interstitial Lung Diseases registry illustrates the 'real world' situation of inpatients and outpatients living with ILD in Germany by providing information on the frequency of all ILD subtypes, comorbidities and diagnostic and treatment patterns.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ Our data show that diagnostic and therapeutic approaches to ILD have changed significantly in recent years. This highlights the fact that the management of ILD is complex and evolving and should be carried out in centres of expertise wherever possible. With the increase in mortality and disability-adjusted life years, efforts are needed for the early detection of ILD.

BACKGROUND

Interstitial lung diseases (ILDs) are a group of more than 200 mainly chronic diseases that affect the lung parenchyma through inflammation and/or fibrosis.[1 2] Diagnosis is made on a multidisciplinary basis using clinical, radiological and pathological aspects of the disease.[2 3] ILDs are rare or very rare.[4] Therefore, information on aetiology, incidence and mortality is often insufficient.[1 5] Evidence to support treatment decisions varies widely between the different ILD subtypes with the emergence of new approaches such as antifibrotic treatment for progressive pulmonary fibrosis (PPF).[6 7] Yet, incidence, mortality and disability-adjusted life years are increasing, according to the 2019 global burden of disease study.[8]

Registry data constitute a valuable source of information to a better understanding of

the complex nature of ILD. In idiopathic pulmonary fibrosis (IPF), several registries have been conducted, such as the German INSIGHTS IPF Registry.[9] Fewer registries include all ILD subtypes, but not always consider the current idiopathic interstitial pneumonias (IIPs) consensus classification based on American Thoracic Society (ATS)/European Respiratory Society (ERS) recommendations.[3] Recently, registry data on PPF have been published: the Canadian Registry for Pulmonary Fibrosis (CARE-PF) included all ILD subtypes and the PROGRESS study (Progressive fibrosing interstitial lung disease: a clinical cohort) all ILD subtypes except IPF. Both registries showed that a progressive course of disease is common in ILD, with 27%–50% of patients developing PPF.[10 11]

The Exploring Clinical and Epidemiological Characteristics of Interstitial Lung Diseases (EXCITING-ILD) registry is a multicentre, non-interventional, prospective and observational disease and outcomes registry conducted by the German Center for Lung Research (DZL) in close collaboration with cross-sectional sites.[5] Here, we provide an overview of the baseline characteristics of the EXCITING-ILD registry. The current work was designed to assess the 'real world' situation of patients living with ILD in Germany, considering the following objectives: (a) frequencies of ILD subtypes, (b) (disease-related) characteristics of patients with ILD, (c) information on diagnostic procedures to identify ILD and (d) management of different (sub)types of ILD.

METHODS

Study design

In the prospective EXCITING-ILD registry, sociodemographic and medical data on all different ILD subtypes were collected. The period during which patients were recruited ran from January 2014 to October 2016, and the final follow-up visit was carried out in July 2021. Incident and prevalent patients with ILD from different healthcare settings, including outpatient, inpatient and academic sites, were included. A total of 32 study centres were involved in the study, not all of which were expert ILD centres. The first visit after enrolment was defined as the baseline. Follow-up visits were performed every 6 months. At baseline and each follow-up visit, the investigators reported the following data, which were entered into the full analysis set: demographic data, information on ILD subtypes, diagnostic procedures, distinct comorbidities, ILD management, as well as outcomes including progression and mortality. All patients with at least one documented post-baseline visit during a median follow-up of 3 years were included in the analyses.[5 12] The full trial protocol has been the subject of publication elsewhere.[5]

Patient representatives were involved in the study design from the beginning and are listed among the coauthors.

Classification of ILD types and subtypes

The EXCITING-ILD registry included patients with almost all ILD subtypes. In order to group them, an established classification based on the recommendations of the ATS and the ERS was used.[2 3 13 14] Online supplemental figure 1 provides an overview of ILD types and subtypes included.

Statistical analysis

Data are presented as absolute numbers and percentages and were analysed descriptively. Mean and SD were used for normally distributed variables. The Wilcoxon signed-rank sum test was used to quantify the difference between groups (multidisciplinary discussion (MDD) vs no MDD). CIs were set at a two-sided level of 95%.[5] Statistical software R (R V.4.1.2) was used for data analysis.

For graphical presentation, line plots were used for the visualisation of the frequencies of surgical lung biopsies and cryobiopsies. Bar plots illustrate the number of patients at each follow-up visit and the frequencies of various drug therapies in IPF.

The ILD-GAP (gender, age, physiology) index was calculated for all patients. The model uses clinical and physiological variables to predict mortality in patients with different forms of ILD. The ILD-GAP index ranges from 0 to 8 and is categorised into three stages (I=0–3, II=4–5, III=6–8) with higher scores indicating a worse prognosis.[15] Forced vital capacity (FVC) and diffusing capacity for carbon monoxide (CO)—single breath (DLCO-SB) values were retrieved from the baseline visit.

For the analyses of treatment in ILD (absolute and relative frequencies of various therapies over the years), the focus was placed on the ILD types and subtypes of special interest. These included 530 patients with IPF, non-specific interstitial pneumonia (NSIP), cryptogenic organising pneumonia (COP), unclassifiable ILD (uILD), sarcoidosis, hypersensitivity pneumonitis (HP), connective tissue disease-associated ILD (CTD-ILD) and drug-induced ILD (DI-ILD). Only long-term therapies were considered, that is, no single doses. In case a patient has more than one entry for a specific drug within 1 year it was only counted once for this year. If a drug was given over a time span of years, it is counted once within each therapy year.

RESULTS

ILD types and subtypes

The EXCITING-ILD registry comprised 601 patients from 32 centres. Almost all different ILD subtypes were included, the frequencies for each ILD type and subtype are shown in table 1. The largest group was IIP with 44.1% (n=265), followed by granulomatous ILD except HP (n=165, 27.5%), HP (n=58, 9.7%) and CTD-ILD (n=43, 7.2%). Overall, the most frequent subtypes were sarcoidosis with 26.6% (n=160) and IPF with 25.3% (n=152).

Table 1 Frequencies of ILD types and subtypes

ILD type	ILD subtype	n	% total	% subtotal
Idiopathic interstitial pneumonia (IIP)		**265**	**44.1**	
	Idiopathic pulmonary fibrosis (IPF)	152	25.3	57.4
	Non-specific interstitial pneumonia (NSIP)	42	7.0	15.8
	Desquamative interstitial pneumonia (DIP)	5	0.8	1.9
	Respiratory bronchiolitis-associated ILD (RB-ILD)	4	0.7	1.5
	Cryptogenic organising pneumonia (COP)	25	4.2	9.4
	Lymphocytic interstitial pneumonia (LIP)	1	0.2	0.4
	Acute interstitial pneumonia (AIP)	1	0.2	0.4
	Pleuropulmonary fibroelastosis (PPFE)	0		
	Others (other IIP)	1	0.2	0.4
	Not classifiable IIP (uILD)	34	5.7	12.8
Granulomatous lung disease (GRAN-ILD)		**165**	**27.5**	
	Sarcoidosis (Sarc)	160	26.6	97.0
	Berylliosis (Beryll)	0		
	Other (eg, involvement in chronic inflammatory liver and gut diseases, except hypersensitivity pneumonitis (EAA)) (other GRAN-ILD)	5	0.8	3.0
Hypersensitivity pneumonitis (HP)		**58**	**9.7**	
	Farmer's lung (F-HP)	8	1.3	13.8
	Bird keepers' lung disease (Bk-HP)	14	2.3	24.1
	Origin unknown (u-HP)	24	4.0	41.4
	Other (other HP)	12	2.0	20.7
Rheumatic and connective tissue diseases with pulmonary involvement (CTD-ILD)		**43**	**7.2**	
	Connective tissue disease (CTD)	22	3.7	51.2
	Vasculitis (Vasc)	6	1.0	14.0
	Rheumatoid arthritis (RA)	15	2.5	34.9
Pneumoconiosis (PNEUCON)		**11**	**1.8**	
	Asbestosis (Asbest)	6	1.0	54.5
	Silicosis (Silico)	5	0.8	45.5
Other forms		**18**	**3.0**	
	Pulmonary lymphangioleiomyomatosis (LAM)	7	1.2	38.9
	Pulmonary Langerhans' cell histiocytosis (PLCH)	2	0.3	11.1
	Pulmonary alveolar proteinosis (PAP)	3	0.5	16.7
	Eosinophilic pneumonia (EP)	6	1.0	33.3
Drug-induced ILD (DI-ILD)		**16**	**2.7**	
Radiotherapy associated ILD (RTX-ILD)		**6**	**1.0**	
Combined pulmonary fibrosis and emphysema (CPFE)		**14**	**2.3**	
Others		**5**	**0.8**	

This table lists absolute and relative frequencies of all ILD types and its subtypes (n=601). The group 'other forms' includes LAM, PLCH, PAP and EP. The last group, 'Others', includes all ILD subtypes not mentioned above, such as IPAF.
ILD, interstitial lung disease; IPAF, interstitial pneumonia with autoimmune features.

Follow-up visits

Follow-up visits were carried out every 6 months. Accordingly, follow-up visit 2 was performed on average 1.03 years after baseline (SD 0.17), follow-up visit 5, 2.55 years after baseline (SD 0.18) and follow-up visit 10, 5.04 years after baseline (SD 0.16). The median follow-up period was 1.5 years (IQR 0.5 years; 2.5 years).

The number of patients per visit decreased over time, with a clear decline after follow-up visit 6, mainly due to deaths and missing visits (online supplemental figure 2).

Baseline characteristics

Most patients (60.7%) were men (online supplemental figure 3). For IPF (n=116, 76.3%) and pneumoconiosis (n=11, 100%), patients were predominantly men. The average age was 64.3 years (SD 14.2). Patients with IPF and combined pulmonary fibrosis and emphysema (CPFE) presented the highest mean age. Familial ILD was present in 3.7% (n=22), and half of the patients were people who currently smoke 8.7% (n=52) and people who used to smoke 44.9% (n=270), respectively. Current smoking showed an accentuation in other forms, including pulmonary Langerhans' cell histiocytosis (PLCH; 22.2%, n=4) and CPFE (28.6%, n=4; online supplemental figure 3).

Reflux was present in 22.5% (n=135) of patients with ILD and pulmonary hypertension in 8.7% (n=52). Pulmonary hypertension was most common in CTD-ILD (n=7, 16.3%) and pneumoconiosis (n=3, 27.3%, online supplemental figure 3).

The mean FVC was 76.4% predicted and the mean DLCO-SB was 54.1% predicted. The lowest FVC value was found in CPFE (65.4% predicted), followed by DI-ILD (68.5% predicted) and HP (69.2% predicted). DLCO-SB was reduced across all ILD subtypes except granulomatous lung disease other than HP. Overall, ILD-GAP index stage I was the most common (45.8%). However, IPF, DI-ILD and CPFE showed a predominance of higher ILD-GAP index stages (online supplemental figure 3). The mean distance in 6 min walking distance test was 371 m (SD 120). The longest distance was recorded in other forms, for example, lymphangioleiomyomatosis or PLCH (530 m, SD 99), and the shortest in CPFE (330 m, n.c., online supplemental figure 3).

Diagnostic work-up

The timely reference point for most time differences was the inclusion date, for example, time to diagnosis was defined as the time from the date of diagnosis to the date of inclusion (time to diagnosis=date of diagnosis−date of inclusion). The mean time to diagnosis was 38.8 months (SD 64.4, online supplemental figure 3) and was significantly shorter when the diagnosis was made in a multidisciplinary team (31.6 vs 49.2 months, p<0.001). The time to diagnosis of granulomatous ILD other than HP was the longest with 63.2 months (SD 92.7). The diagnostic work-up included bronchoscopy in 75.7% of the cases, most commonly with bronchoalveolar lavage (68.2%). Bronchoscopy was least frequently performed in pneumoconiosis (n=3, 27.3%), CTD-ILD (n=29, 67.4%), DI-ILD (n=11, 68.8%) and CPFE (n=9, 64.3%).

Cryobiopsies were performed in 32.3% and endobronchial ultrasound-guided transbronchial needle aspiration in 23.1%, the latter mainly for granulomatous ILD (49.6%) other than HP. Over time in the registry, a decrease in the frequency of surgical lung biopsies could be registered (<2010: 10.2%, 2016: 0.4%, figure 1). Meanwhile, the proportion of cryobiopsies increased notably (<2010: 5.9%, 2016: 10.4%; figure 1).

Treatment

The following ILD treatment analyses were performed for a selection of ILD of special interest: IPF, iNSIP, COP, uILD, sarcoidosis, HP, CTD-ILD and DI-ILD (n=530). Of these, 181 (2019) to 403 (2016) patients received treatment in the years under review.

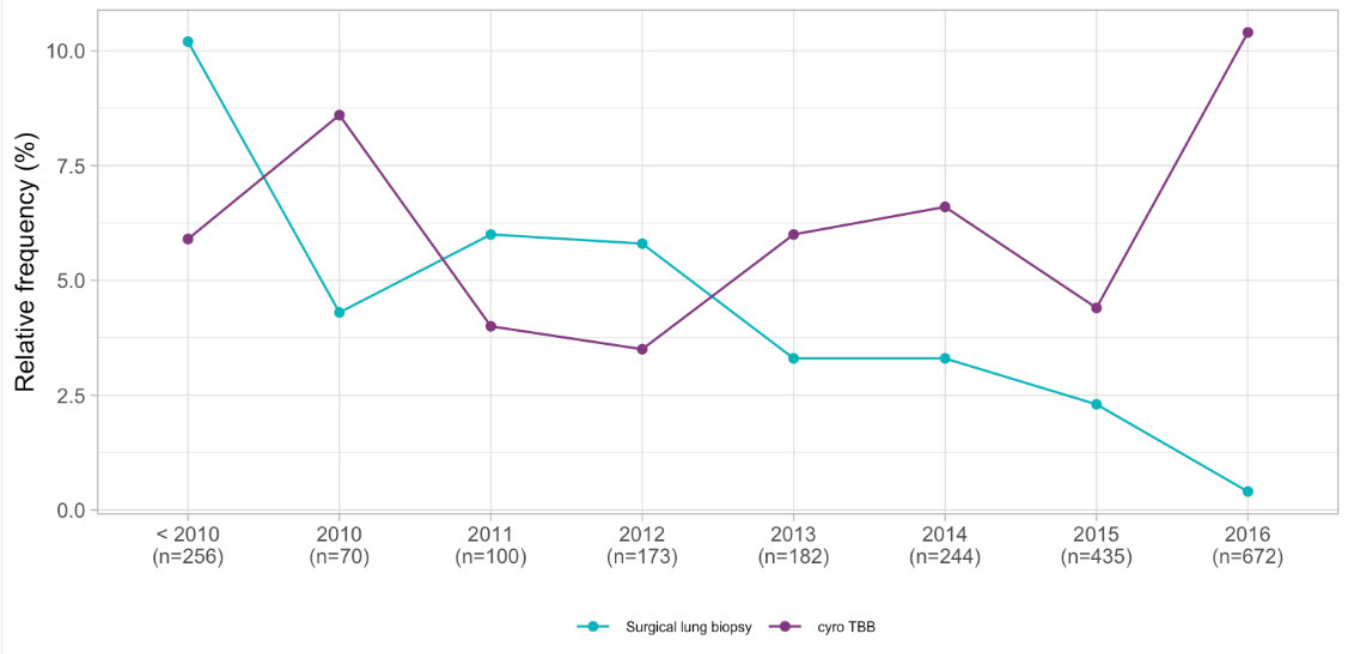

Figure 1 Frequencies of surgical lung biopsy (blue) and cryo-TBB grouped by year. Relative frequencies were computed based on the total number of diagnostic procedures (n) in the corresponding year. TBB, transbronchial biopsy.

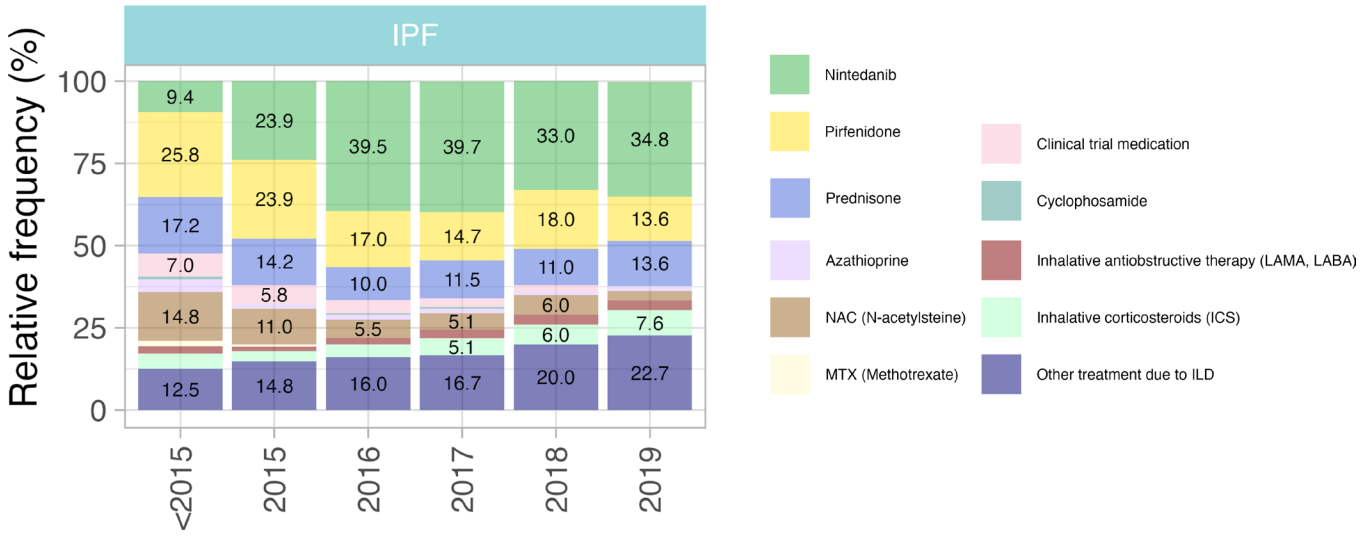

Figure 2 Frequencies of drug therapies in IPF grouped by year. Only therapies with a relative frequency of more than 5% in the corresponding year were labelled. ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; LABA, long-acting beta2-agonist; LAMA, long-acting muscarinic antagonist.

At baseline, the most frequently used drug was prednisone (39.1%, n=169). Other recorded immunosuppressants were azathioprine (n=27, 6.2%) and methotrexate (n=19, 4.4%). Less commonly used immunosuppressants were cyclophosphamide with 1.6% (n=7), rituximab with 0.9% (n=4), mycophenolate mofetil (MMF) with 0.7% (n=3) and TNF-alpha blockers with 0.2% (n=1). The antifibrotics nintedanib and pirfenidone were applied in 2.8% (n=12) and 7.6% (n=33), respectively. Between 0.5% and 2.1% received a clinical trial medication, with a maximum before 2015. N-acetylcysteine (NAC) was used in 6.7% (n=29), inhaled antiobstructive therapy (long-acting muscarinic antagonist, long-acting beta2-agonist) in 6.7% (n=29), inhaled corticosteroids in 7.9% (n=34) and other treatments such as long-term oxygen therapy (LTOT) or physiotherapy in 13.0% (n=56).

In IPF, the treatment regime has changed significantly over the period of the registry (figure 2). The proportion of patients receiving antifibrotic treatment with either nintedanib or pirfenidone increased from 35.2% before 2015 to 48.4% in 2019. Before 2015, the use of pirfenidone predominated (25.8% vs nintedanib 9.4%), in 2015 the proportion was 23.9% each, and since 2016 nintedanib has been used more frequently (13.6% vs pirfenidone 34.8%). Over the same period, the use of prednisone decreased, but remained at 13.6% in 2019. The same applied to NAC, which remained at 3.0% in 2019 (figure 2). Other treatments increased over time, mainly due to the growing use of LTOT (<2015 640.7%, 2019 85.0%). Treatment regimens for non-IPF ILD are presented in online supplemental figure 4.

DISCUSSION

The present study is based on data from the EXCITING-ILD registry, a registry including prevalent and incident patients of all ILD subtypes, reporting epidemiological data, baseline characteristics and information on diagnostic work-up and therapeutic approach of these rare or very rare diseases.

Incidence and disability-adjusted life years increase in ILD over time.[8] In addition, ILD often has a progressive disease course, leading to poor disease outcomes and high mortality.[12] In fact, mortality is largely caused by ILD directly, especially in fibrotic ILD.[12] Reports on registries covering all different ILD according to the current classifications are sparse. Therefore, general comparability between the existing registries is difficult. In the EXCITING-ILD registry, ILD types and subtypes were classified according to the 2013 ATS/ERS International Multidisciplinary Consensus Classification of Idiopathic Interstitial Pneumonias.[3] In addition, patients from both inpatient and outpatient settings were included to avoid underrepresentation of specific ILD subtypes such as sarcoidosis and to provide an as accurate picture as possible of the 'real world' situation of patients with ILD in Germany.[5]

In the total of 601 patients, the most common subtypes were sarcoidosis (26.6%) and IPF (25.3%). Many other registries also reported sarcoidosis and IPF as the most common ILD subtypes. This was shown both by data from Belgium from around the 2000s with 31% sarcoidosis and 20% IPF[16 17] and by data from the German 'Fibrosis registry' from 2003 with 44.7% sarcoidosis and 27.0% IPF.[18] Together, these two subtypes usually account for more than 50% of all ILDs.[5 19] Apart from PPFE and berylliosis, all other ILD subtypes are represented with sufficient numbers. Our data showed a male predominance of 60.7% of all represented ILD, as similarly reported in one of the first ILD registries between 1988 and 1990,[20] and partially explained by the included ILD subtypes. Male sex and elderly age were detected to be characteristic for IPF and exposure-related ILD, smoking for PLCH and CPFE.

The baseline pulmonary function testing values have already shown a mild impairment. With a mean FVC of 76.4% predicted and a mean DLCO-SB of 54.1% predicted, current results are comparable to other registries including all ILD entities such as the CARE-PF registry.[21] This finding is particularly relevant as in our previous work, the most important predictor of progression was a reduced baseline FVC, which also associated with older age.[12] Higher GAP indices, which use clinical and physiological variables to predict mortality in patients with different forms of ILD,[15] were found in IPF, consistent with the known poor prognosis of the disease.[6] In addition, previous analyses showed that patients with higher GAP indices were more likely to be hospitalised.[22]

Within the EXCITING-ILD registry, the mean time to diagnosis was 38.8 months. Based on a survey of barriers to timely ILD diagnosis in 600 participants, Cosgrove *et al* reported ≥1 misdiagnosis in 55% and ≥2 misdiagnoses in 38% prior to the current diagnosis. Common misdiagnoses were asthma, pneumonia and bronchitis. Time to diagnosis exceeded 2 years in 34% after initial misdiagnosis.[23] Delaying appropriate treatment due to the common non-specific symptoms, the complex diagnostic procedure and limited access to specialised ILD centres showed a detrimental effect on quality of life.[23] One way to address this is to require multidisciplinary discussion for the diagnosis of ILD, as suggested in the recent guidelines.[6] Accordingly, the current registry study showed that the time to diagnosis was significantly shorter when the diagnosis was made in an MDD. In the light of increased disease mortality despite new therapeutic options, current efforts are directed towards early detection of fibrotic ILD.[24]

44.3% of patients had a lung biopsy as part of the diagnostic work-up. This is comparable to Spanish registry data, which reported a biopsy-confirmed diagnosis in 40.1% of 744 patients.[25] Our data showed a remarkable shift from surgical lung biopsy (SLB) to transbronchial lung cryobiopsy (TBLC) within this regard. Troy et al. in 2020 showed a high concordance in terms of histopathological and MDD diagnosis between TBLC and SLB.[26]

In recent years, therapeutic approaches in ILD have changed significantly. Our data illustrate this through the example of IPF. The use of the two antifibrotics nintedanib and pirfenidone increased from 35.2% before 2015 to 48.4% in 2019. After the 'Clinical Studies Assessing Pirfenidone in idiopathic pulmonary fibrosis: Research of Efficacy and Safety Outcomes' programme led to the approval of pirfenidone in Germany in 2011,[27] treatment with pirfenidone predominated in the EXCITING registry until 2015. Based on the "Nintedanib in Idiopathic Pulmonary Fibrosis: The Phase 3 INPULSIS™ Trials",[28] nintedanib followed in 2015 and, according to registry data from 2016, a shift in prescribing towards nintedanib continued. Surprisingly, although the use of prednisone has decreased notably, it remained at 13.6% in 2019. The "Prednisone, Azathioprine, and *N*-Acetylcysteine: A Study That Evaluates Response in Idiopathic Pulmonary Fibrosis" trial marked a turning point, showing excess mortality in patients with IPF on corticosteroid therapy.[29] This highlights once again the fact that treating ILD is complex and evolving and should be carried out in centres of expertise wherever possible.

Only a selection of comorbidities was registered in the current registry. Consistent with previous data, reflux was the most commonly registered comorbidity in ILD.[21] Another commonly seen comorbidity was pulmonary hypertension. This was found to be most common in CTD-ILD and in pneumoconiosis. The overall frequency was in line with the data for uILD[30] and guidelines recommending screening for pulmonary hypertension, such as in systemic sclerosis-associated ILD.[31]

The EXCITING-ILD registry was designed to assess the 'real world' situation of patients living with ILD in Germany. Its particular strengths are the long observation period and the multicentre approach, which takes into account different healthcare settings. However, there are a number of limitations. Different healthcare settings bring with them varying levels of expertise in ILD. Therefore, clinical decisions, for example, regarding therapy, may differ between treating physicians. Furthermore, it is not possible to distinguish between incident and prevalent patients, as both were included. As a result, the characteristics at the time of diagnosis may not be fully reflected. Another limitation is the potential for time bias, as the data collection occurred over a limited timeframe, which may not fully represent long-term trends; additionally, referral bias could have influenced the sample, as participants were selected from specific referral sources, potentially limiting the generalisability of the findings. The EXCITING-ILD registry did not include information on CT scans. In addition, comorbidities were not fully captured.[5 12 22]

CONCLUSION

In a cohort of 601 patients from different healthcare facilities, the EXCITING-ILD registry described the frequency of all ILD subtypes, moreover the lung function impairment present at baseline and selected comorbidities. Furthermore, the current registry highlights change in diagnostic procedures and treatment of ILD over the course of the registry.

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