Real-life characteristics and management of patients with fibrosing interstitial lung disease: INSIGHTS-ILD registry

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This study aims to obtain real-life data on the characteristics and treatment trajectories of patients with fibrosing interstitial lung disease (fILD).

INSIGHTS-ILD is an ongoing prospective registry for fILDs conducted at 32 expert sites across Germany. fILD patients are eligible if single breath diffusion capacity of the lung for carbon monoxide (DLCO) ≤80% predicted, interstitial lung fibrosis >10% on HRCT, and are on any active anti-inflammatory and/or antifibrotic therapy. Criteria of progression were not mandatory for inclusion. Patients with IPF are excluded. Study registration: DRKS00027389, EMA RWD Catalogue 1000000161.

Characteristics of the 655 patients in the interim analysis are presented in Figure 1. The most frequently reported ILD entities were fibrotic hypersensitivity pneumonitis (31.2%), fibrotic idiopathic interstitial lung disease (22.0%), rheumatoid arthritis and connective tissue disease ILDs (13.0%), and unclassifiable fILD (13.0%). For baseline characteristics see Table 1. Current treatment included oral steroids (62.6%), antifibrotic therapy (50.5%), azathioprine (14.4%), MTX (10.2%) and MMF (11.1%). Mean quality of life on the 0-100 visual analogue scale was 58±19 points.

Patients with vs. without antifibrotic therapy had a higher prevalence of INBUILD criteria for progression (72.7% vs. 40.1%).

The patient characteristics in this registry closely resemble those observed in randomized controlled trials and non-interventional studies of fILD, with individuals on antifibrotic therapy displaying a more severe disease profile.

Table 1. Demographic and clinical characteristics, total and by treatment strategy at baseline

Category	Antifibrotic Therapy (n=331)	No Antifibrotic Therapy (n=324)	All Patients (n=655)
Age (years)	67.7 (10.5), 69.0	64.1 (12.7), 66.0	65.9 (11.7), 68.0
Gender – Female	131 (39.6%)	167 (51.5%)	298 (45.5%)
Gender – Male	200 (60.4%)	157 (48.5%)	357 (54.5%)
BMI (kg/m²) - < 18.5	9 (2.7%)	6 (1.9%)	15 (2.3%)
BMI (kg/m²) - [18.5; 25]	124 (37.5%)	104 (32.1%)	228 (34.8%)
BMI (kg/m²) - [25; 30]	118 (35.6%)	111 (34.3%)	229 (35.0%)
BMI (kg/m²) - ≥ 30	80 (24.2%)	103 (31.8%)	183 (27.9%)
Smoking – Never	135 (42.7%)	143 (47.5%)	278 (45.1%)
Smoking – Former	177 (56.0%)	145 (48.2%)	322 (52.2%)
Smoking – Current	4 (1.3%)	13 (4.3%)	17 (2.8%)
Comorbidities	1.8 (1.6), 2.0	1.6 (1.4), 1.0	1.7 (1.5), 1.0
Duration of symptoms (years)	6.8 (6.2), 4.9	6.3 (7.8), 3.9	6.5 (7.1), 4.5
Age at symptom onset (years)	60.8 (12.1), 62.0	57.8 (15.0), 61.0	59.3 (13.7), 61.0
Age at diagnosis (years)	62.6 (11.8), 64.0	59.4 (14.7), 62.0	61.0 (13.4), 63.0
6-minute walk distance (m)	338.2 (114.9), 360.0	401.6 (118.8), 410.0	365.1 (120.6), 372.5
Borg dyspnea index at rest	1.2 (1.9), 0.0	1.3 (2.1), 0.0	1.2 (2.0), 0.0
Borg dyspnea index after exercise	4.9 (2.5), 5.0	4.8 (2.4), 5.0	4.9 (2.4), 5.0
Environmental influences	173 (63.8%)	129 (53.5%)	302 (59.0%)
Gastroesophageal reflux disease	51 (16.5%)	51 (17.3%)	102 (16.9%)
Familial clustering	28 (12.8%)	22 (10.0%)	50 (11.4%)
Medication exposure	36 (14.3%)	19 (8.3%)	55 (11.5%)
Patients fulfilling at least one of the INBUILD progression Criteria	189 (72.7%)	109 (40.1%)	298 (56.0%)
Patients fulfilling none of the INBUILD progression criteria	71 (27.3%)	163 (59.9%)	234 (44.0%)

Values are mean (standard deviation), median or n (%)

Table 2. Treatment at baseline

Medication	Antifibrotic Therapy (n=331)	No Antifibrotic Therapy (n=324)	All Patients (n=655)
Nintedanib	318 (96.1%)	0 (0.0%)	318 (48.5%)
Pirfenidone	14 (4.2%)	0 (0.0%)	14 (2.1%)
Prednisone/Prednisolone	175 (52.9%)	235 (72.5%)	410 (62.6%)
Other Steroid	0 (0.0%)	3 (0.9%)	3 (0.5%)
Azathioprine	27 (8.2%)	67 (20.7%)	94 (14.4%)
Cyclophosphamide IV	4 (1.2%)	9 (2.8%)	13 (2.0%)
Cyclophosphamide Oral	0 (0.0%)	0 (0.0%)	0 (0.0%)
Mycophenolate Mofetil (MMF)	31 (9.4%)	42 (13.0%)	73 (11.1%)
Rituximab	15 (4.5%)	33 (10.2%)	48 (7.3%)
Methotrexate (MTX)	24 (7.3%)	43 (13.3%)	67 (10.2%)
Other	14 (4.2%)	36 (11.1%)	50 (7.6%)

Values are n (%). Multiple medications were possible.

Table 3. Patient characteristics by etiology at baseline

Parameter	FIIP (n=133)	HP (n=190)	Rheumatoid Arthritis (n=80)	Systemic Sclerosis (n=66)
Age (years)	68.0 (11.3), 70.0	66.6 (10.4), 68.0	70.3 (9.7), 70.0	62.6 (10.9), 64.0
Sex, female	47 (35.3%)	91 (47.9%)	37 (46.2%)	38 (57.6%)
Sex, male	86 (64.7%)	99 (52.1%)	43 (53.8%)	28 (42.4%)
BMI (kg/m²)	27.3 (5.0), 27.1	28.5 (5.7), 27.5	27.5 (5.0), 26.9	24.9 (4.3), 23.7
Smoking never	57 (44.2%)	86 (46.0%)	25 (32.1%)	29 (49.2%)
Smoking past	69 (53.5%)	99 (52.9%)	48 (61.5%)	27 (45.8%)
Smoking current	3 (2.3%)	2 (1.1%)	5 (6.4%)	3 (5.1%)
Comorbidities	1.8 (1.5), 1.0	1.7 (1.5), 1.0	2.0 (1.8), 2.0	1.6 (1.5), 1.0
Symptom duration at entry (yrs)	5.3 (5.6), 3.7	7.9 (8.5), 5.1	6.0 (5.8), 4.3	6.8 (6.4), 4.3
Age symptom begin (yrs)	62.6 (12.4), 63.0	58.5 (13.9), 59.0	64.5 (10.1), 65.0	55.9 (12.3), 57.0
Age at diagnosis (yrs)	64.1 (12.1), 66.0	61.0 (13.5), 63.0	64.9 (10.8), 66.0	56.8 (12.3), 57.5
6-minutes walking distance (m)	356 (131), 360	354 (119), 361	326 (134), 330	416 (111), 426
Borg dyspnoea index resting	1.5 (2.3), 0.0	1.4 (2.3), 0.0	1.1 (1.8), 0.0	1.4 (1.8), 1.0
Borg dyspnoea index exercise	4.9 (2.6), 5.0	5.2 (2.3), 5.0	4.9 (2.8), 5.0	4.2 (2.3), 5.0
Environmental influences	46 (46.0%)	142 (81.6%)	29 (48.3%)	25 (58.1%)
Gastroesophageal reflux	18 (15.0%)	26 (14.1%)	7 (9.5%)	22 (37.3%)
Familial clustering	9 (9.2%)	16 (12.2%)	7 (12.5%)	5 (12.2%)
Medication exposure	11 (11.2%)	14 (9.3%)	15 (25.4%)	3 (7.5%)

Values are mean (standard deviation), median or n (%)

Table 4. Lung function, by etiology

Parameter	FIIP (n=133)	HP (n=190)	Rheumatoid Arthritis (n=80)	Systemic Sclerosis (n=66)
Total Lung Capacity (TLC) (% pred.)	69.1 (17.4), 68.3	65.3 (16.5), 64.2	69.1 (16.6), 68.3	71.2 (17.3), 70.1
Inspiratory Vital Capacity (IVC) (% pred.)	72.8 (24.2), 72.4	65.7 (17.9), 65.8	71.5 (18.2), 70.9	71.2 (22.3), 72.0
Forced Vital Capacity (FVC) (% pred.)	71.8 (20.1), 71.5	65.2 (17.5), 65.2	72.4 (19.6), 70.0	72.5 (22.2), 72.3
Forced Expiratory Volume in 1 second (FEV1) (% pred.)	76.6 (20.2), 75.4	69.4 (18.1), 68.2	76.9 (17.8), 74.4	74.8 (20.7), 74.6
FEV1/FVC Ratio (% pred.)	112.5 (10.1), 112.5	112.8 (8.8), 113.5	113.2 (9.6), 113.4	110.6 (10.4), 110.4
Diffusing Capacity for CO (DLCO) (% pred.)	34.0 (16.2), 30.3	31.2 (14.4), 26.9	34.0 (14.6), 32.1	35.7 (18.1), 32.3

Values are mean (standard deviation), median

Table 5. Treatment in inclusion, by etiology

Treatment	FIIP (n=133)	HP (n=190)	Rheumatoid Arthritis (n=80)	Systemic Sclerosis (n=66)
Nintedanib	66 (49.6%)	100 (52.6%)	40 (50.0%)	33 (50.0%)
Pirfenidone	5 (3.8%)	3 (1.6%)	2 (2.5%)	0 (0.0%)
Prednisone/Prednisolone	82 (61.7%)	136 (71.6%)	45 (56.2%)	27 (40.9%)
Other Steroid	1 (0.8%)	1 (0.5%)	0 (0.0%)	0 (0.0%)
Azathioprine	16 (12.0%)	34 (17.9%)	4 (5.0%)	10 (15.2%)
Cyclophosphamide IV	0 (0.0%)	2 (1.1%)	0 (0.0%)	6 (9.1%)
Cyclophosphamide Oral	0 (0.0%)	0 (0.0%)	0 (0.0%)	0 (0.0%)
MMF	9 (6.8%)	15 (7.9%)	5 (6.2%)	22 (33.3%)
Rituximab	4 (3.0%)	1 (0.5%)	20 (25.0%)	4 (6.1%)
MTX	10 (7.5%)	10 (5.3%)	24 (30.0%)	9 (13.6%)
Other	5 (3.8%)	3 (1.6%)	19 (23.8%)	7 (10.6%)

Values are n (%). Multiple medications were possible

Figure 1. Diagnoses at baseline

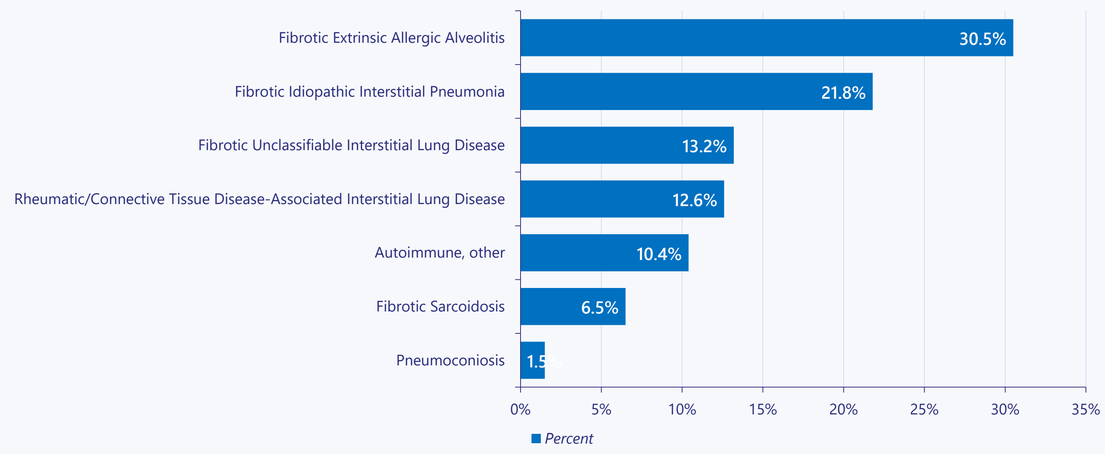


Figure 2. Symptoms at baseline

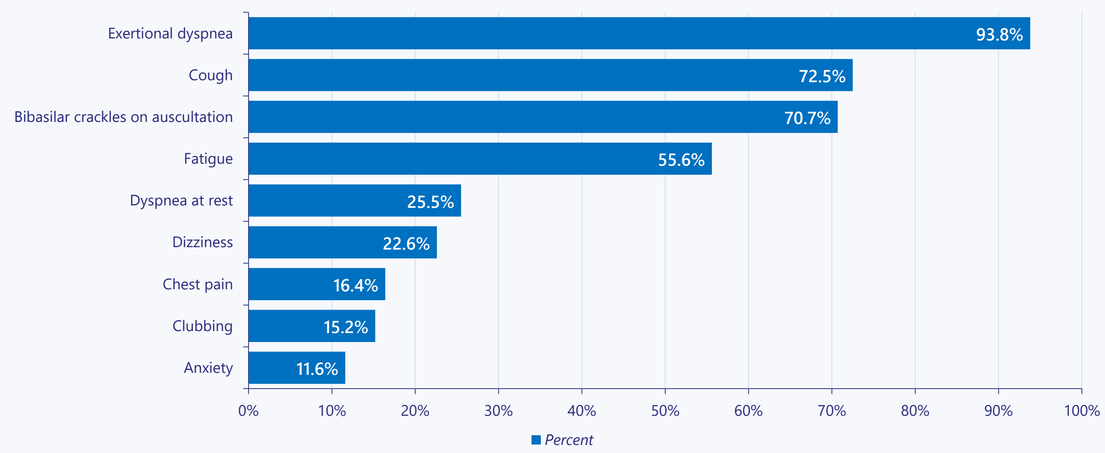
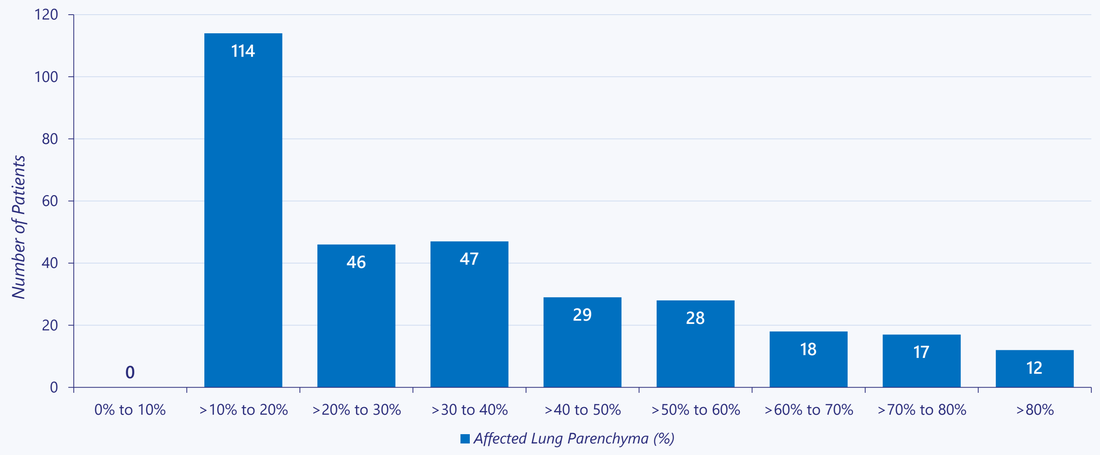


Figure 3. Affected lung parenchyma by percentage range in HRCT



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