The 26th European Respiratory Society International Congress, (ERS) – the largest respiratory meeting in the world – was held over the 3rd – 7th of September in London, United Kingdom. International experts discussed new data with regards to interstitial lung diseases (ILD) and idiopathic pulmonary fibrosis (IPF).

**ERS 2016 Congress Highlights – Interstitial Lung Disease (ILD)**

London, UK
September 3rd – 7th 2016

The preparation of this PDF was sponsored by Boehringer Ingelheim International GmbH. ERS was neither author nor reviewer of the content.
Diagnosis and multidisciplinary discussion for ILD

New findings around the diagnosis of ILD and IPF

Early diagnosis and treatment of IPF

New data from the EMPIRE-IPF¹ registry show that early diagnosis and treatment of IPF matters in the prognosis of patients with IPF.² The 566 patients were categorized by time from first symptoms to diagnosis below or above 1 year. Compared to the later diagnosed group (n=183), the earlier diagnosed group (n=383) showed better median survival from the diagnosis up to 84 months (63.1 vs 43.9 months; p=0.018) and a higher VC at the time of diagnosis (82.9% of predicted vs 75.8% of predicted; p=0.008). There was no difference in rate of VC decline.

Data from other registries which were presented at the congress also emphasized the need for improved, earlier diagnosis of ILD and IPF.

• Results from the British BTS IPF registry showed that 47% of the 767 patients with IPF had already had symptoms for more than 24 months before presentation³

Diagnosis of UIP on HRCT- Impact of the 2011 IPF diagnostic guidelines

The investigators asked two Canadian general pulmonologists to review HRCTs of patients with ILD before and after applying current IPF diagnostic guidelines⁴ to identify UIP pattern on HRCT.⁵ Application of the guidelines led to improved agreement for possible UIP (from 66% to 72%) and inconsistent UIP (from 78% to 88%), but not for definite UIP (from 75% to 78%). This is of some concern, since according to the 2011 diagnostic guidelines, the only way to diagnose IPF without conducting a lung biopsy is a definite UIP pattern on HRCT.
Surgical lung biopsy (SLB) for ILD and associated mortality rates

Analysis of Hospital Episodes Statistics data from 1997-2008 showed the associated mortality rates of SLB for the diagnosis of ILD in England (n= 2937). The researchers noted that the number of biopsies increased over time and identified the following risk factors for mortality:

- Male sex
- Increasing age
- Increasing co-morbidity
- Open surgery

The most common cause of death for all cases was interstitial lung disease.

Frequency of diagnostic procedures for patients with ILD in Germany

The German EXCITING-ILD registry (n=201) reported the following frequency of diagnostic procedures for patients with ILD:

- CT: 91%
- HRCT: 49%
- PFT: 88%
- BAL: 75%
- SLB: 24%
- MDT: 58%

CT=Computed Tomography; HRCT=High-Resolution CT; PFT=Pulmonary Function Test; BAL=Bronchoalveolar lavage; SLB=Surgical Lung Biopsy; MDT=Multidisciplinary Team
MDT is a best practice for the diagnosis of ILD

MDT as best practice for the diagnosis of ILD

The importance of a multi-disciplinary team (MDT) as best practice for the diagnosis of ILD was highlighted by a Spanish investigation. The study evaluated the diagnostic steps taken for all patients assessed in the Bellvitge University Hospital ILD Unit during 2014 (n=158). Diagnosis was obtained by the MDT, evaluating the following parameters stepwise, until a confident diagnosis was reached:

1. Detailed patient history (including antibodies and family history)
2. HRCT evaluation by two independent radiologists
3. Pathology
4. MDT committee discussion

Nearly all patients could be diagnosed and in 23 cases (14.6%), committee discussion determined the diagnosis. In addition, the initial diagnosis of 18 out of 91 cases (19.8 %) had to be modified after Multidisciplinary Team Discussion (MDD).

The importance of MDT for diagnosis was also demonstrated by results from the British BTS-IPF registry. The investigators found that 90% of cases diagnosed with IPF (out of a total of 767 patients) were reviewed by MDT.

Patients with IPF are often exposed to occupational and domestic hazards

Interim results from the PROOF registry (Belgium and Luxembourg) show that patients with IPF (n=175) might be exposed to occupational and/or domestic hazards more often than generally assumed. The authors highlight that a history of exposure makes IPF diagnosis more difficult and that, therefore, MDT is more important.
Real world experience with anti-fibrotic treatments

Researchers from the ILD clinic at Hammersmith Hospital (UK) reviewed their data collected on patients with IPF treated with either pirfenidone (4/2013-2/2016) or nintedanib (2/2015-2/2016). Of the 36 patients, those treated with nintedanib were more likely to tolerate the maximum dose and to remain on treatment compared to patients treated with pirfenidone.

Nintedanib for IPF treatment in the real world

Several studies evaluated the safety and efficacy profile of nintedanib in real-life clinical settings. All of them confirmed nintedanib's safety profile for the treatment of IPF and reported a slowed decline in FVC % pred. over time for patients treated with nintedanib. Though longer observation periods are needed, these data agree with the previously published data.

Experience with nintedanib in Greece

Kontou et al. performed a study within the scope of a named patient use (NPU) program in Greece from 11/2014 to 1/2016. 80 patients were included regardless of the severity of their IPF disease. Treatment discontinuation took place in 23 of these cases (29%), 14 of which were due to ADRs and intolerance of low dosage. Treatment with nintedanib was rated tolerable and safe, with the most common ADR having been diarrhea (reported by 46% of patients).

German compassionate use program for nintedanib

Besides a favorable safety profile, Bonella et al. confirmed that treatment with nintedanib is associated with stabilization of the FVC % pred. in most patients within the German compassionate use program (CPU) including 62 patients. 77% of the patients had switched from treatment with pirfenidone, in most cases as a result of disease progression. Treatment with nintedanib led to stabilization of lung function (mean FVC decline 3±1%) in 63% of the patients. While 63% of patients reported diarrhea, only 10% discontinued treatment.

Nintedanib in Germany: experience from 60 patients

Brunnemer et al. also confirmed the safety of nintedanib in 60 patients. They reported two cases of discontinuation, and diarrhea was the most common ADR (19%). Lung function measured by FVC % pred. and DLCO % pred. (70% and 41% at baseline) declined by 4.4% and 1.2%, respectively, over a mean drug exposure time of 11 months. 33% of patients discontinued treatment.

UK experience with nintedanib

Data from early clinical experience with nintedanib in IPF from three UK tertiary referral centers, and patients treated between 12/2014 - 01/2016 were analyzed retrospectively. This study, led by Toellner, included 188 patients, who either had an initial FVC of >80% pred. (50%; n=94) or who experienced intolerable AEs (22%, n=42) or disease progression during pirfenidone treatment (2%, n=4), or who refused pirfenidone due to its AE profile (17%, n=32). Although less frequent than in the INPULSIS trials, diarrhea was reported as the most common AE (22%) and 14% (n=27) of the patients discontinued treatment due to AEs.
Real-world experience with pirfenidone in IPF

Efficacy of pirfenidone in patients with mild-to-moderate disease

A retrospective study of 99 patients diagnosed with mild-to-moderate IPF who started on pirfenidone analyzed changes in lung function from start of treatment until 12 months of follow up at a 3-month interval. This analysis described a stabilizing effect of pirfenidone on lung function decline in 99 patients with mild-to-moderate IPF.16

A large number of patients remained stable in terms of lung function (defined as change in FVC % pred. between 5 and -5%) and 14.1% even showed an increase.

Giudice et al. also investigated IPF disease progression before and during pirfenidone treatment.17 Between 2011 and 2016, 20 patients with mild-to-moderate IPF were examined with regards to pulmonary function before and during treatment with pirfenidone.

After an initial decline in lung function before initiation of treatment, disease progression (reflected by FVC % pred.) remained stable after introduction of pirfenidone.
Efficacy of pirfenidone in patients with advanced disease

Evidence of pirfenidone efficacy in patients with advanced disease (FVC ≤ 50% and/or DLco ≤ 35%) was found in a small study conducted in Belgium. Of the 23 initially included patients, the 16 patients remaining on treatment showed a significantly reduced decline in lung function after the first six months of therapy (single-sided paired T-test), suggesting a benefit from pirfenidone for patients with advanced IPF.

Anorexia might be a risk factor for treatment discontinuation with pirfenidone

A retrospective study by Teraoka et al. (01/2011 to 01/2016) investigated reasons of intolerance to pirfenidone. Out of the 76 patients studied, 19 discontinued treatment with pirfenidone. The most common AE leading to discontinuation was anorexia (11 out of 76 patients). Compared to the control group of patients remaining on pirfenidone for over one month, the discontinuation group took prophylactic dopamine receptor antagonists less frequently (38% vs 9%, p=0.04).
Long-term data on approved treatments for IPF

Long-term data on nintedanib from INPULSIS®-ON

The long-term efficacy and safety of nintedanib is being assessed based on data from the INPULSIS®-ON open label study. The interim analysis presented at this year’s ERS is based on data collected until October 2015 and includes 734 patients (430 continuing nintedanib; 304 initiating nintedanib). Results show that the treatment effect of nintedanib on slowing disease progression persists for 3 years (similar decline in FVC % pred. in patients in INPULSIS®-ON compared to patients on nintedanib in INPULSIS®). Long-term nintedanib treatment (up to 51 months) had a manageable safety and tolerability profile, with no new safety signals identified.

Change in FVC in INPULSIS® and INPULSIS®-ON

<table>
<thead>
<tr>
<th></th>
<th>Nintedanib Baseline to Week 52 (n=519)</th>
<th>Placebo Baseline to Week 52 (n=345)</th>
<th>INPULSIS®</th>
<th>Mean (SEM) observed change in FVC (mL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continuing nintedanib Baseline to Week 48 (n=352)</td>
<td>-88.9</td>
<td>-203.0</td>
<td>-250</td>
<td>-200</td>
</tr>
<tr>
<td>Week 48 to Week 96 (n=287)</td>
<td>-96.4</td>
<td>-124.3</td>
<td>-250</td>
<td>-200</td>
</tr>
<tr>
<td>Initiating nintedanib Baseline to Week 48 (n=233)</td>
<td>-73.1</td>
<td>-99.6</td>
<td>-250</td>
<td>-200</td>
</tr>
<tr>
<td>Week 48 to Week 96 (n=175)</td>
<td>-99.6</td>
<td>-250</td>
<td>-200</td>
<td>-150</td>
</tr>
</tbody>
</table>
Long term data on pirfenidone from RECAP
The long-term safety profile of pirfenidone was assessed in the RECAP21 (NCT00662038) open label extension study (n=1058). The study was open to patients with IPF who had completed one of the ASCEND22 or CAPACITY23 Phase 3 trials. The final analysis based on data collected from 09/2008 to 06/2015 shows a safety profile for pirfenidone consistent with previous analyses and without new safety signals.24

In addition to these results, the pooled analysis of safety data from ASCEND/CAPACITY22,23 and RECAP (data cut 06/2015, n=1216) are in line with results obtained from RECAP alone, as well as with previous analyses.25

<table>
<thead>
<tr>
<th></th>
<th>RECAP final analysis</th>
<th>ASCEND/CAPACITY/RECAP pooled</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mean dose</strong></td>
<td>2091.1 mg/d</td>
<td>2306 mg/d</td>
</tr>
<tr>
<td><strong>Median exposure</strong></td>
<td>7.3 months (88 weeks) (2482 patient-exposure years)</td>
<td>25.9 months (3366 patient-exposure years)</td>
</tr>
<tr>
<td><strong>Treatment emergent AE</strong></td>
<td>Occurred in 98% of patients</td>
<td>54% serious TEAE: • 21.7% IPF • 8.5% Pneumonia</td>
</tr>
<tr>
<td><strong>Discontinuation</strong></td>
<td>• Caused by TEAE in 42% of patients • Main cause: IPF (17%)</td>
<td>• Caused by TEAE in 45% of patients • Main cause: IPF (15.9%)</td>
</tr>
<tr>
<td><strong>Deaths</strong></td>
<td>• Occurred in 22% of patients • Most common cause: IPF (13%)</td>
<td>82.6 months</td>
</tr>
</tbody>
</table>
Subgroup analyses for nintedanib and pirfenidone

Nintedanib in IPF: Pooled post-hoc analyses of the INPULSIS® trials

No effect of baseline CPI on disease progression during nintedanib treatment

The effect of nintedanib on FVC decline was compared for different composite physiologic index (CPI) subgroups. CPI might predict the extent of fibrosis better than individual parameters for lung function (with a higher CPI predicting a higher extent of fibrosis), and is calculated using DLco, FVC and FEV1.

Patients with CPI ≤45 vs >45 and ≤55 vs >55 at baseline were compared on the primary and key secondary endpoints. As demonstrated in previous analyses, nintedanib reduced the annual rate of decline in FVC and no significant difference in the effect of nintedanib between the subgroups could be identified.

However, in the group of patients with CPI>45 at baseline (compared to ≤45 at baseline)

- More patients experienced an acute IPF exacerbation (but no significant difference in the treatment effect of nintedanib between groups was detected)
- Patients reported a greater increase in SGRQ total score (equaling a worsening of quality of life) and these patients also displayed a more pronounced treatment effect with nintedanib

<table>
<thead>
<tr>
<th>CPI ≤45</th>
<th>CPI &gt;45</th>
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<tbody>
<tr>
<td>n=278</td>
<td>n=360</td>
</tr>
<tr>
<td>-108.4</td>
<td>-118.8</td>
</tr>
<tr>
<td>Δ101.5 mL (95% CI: 51.4, 151.6)</td>
<td>Δ116.4 mL (95% CI: 69.3, 163.5)</td>
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</tbody>
</table>

<table>
<thead>
<tr>
<th>CPI ≤55</th>
<th>CPI &gt;55</th>
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</thead>
<tbody>
<tr>
<td>n=509</td>
<td>n=320</td>
</tr>
<tr>
<td>-104.3</td>
<td>-156.1</td>
</tr>
<tr>
<td>Δ104.5 mL (95% CI: 67.5, 141.5)</td>
<td>Δ123.8 mL (95% CI: 36.6, 211.0)</td>
</tr>
</tbody>
</table>

Annual rate of decline in FVC by CPI ≤ and >45 at baseline

Annual rate of decline in FVC by CPI ≤ and >55 at baseline

Treatment-by-time-by-subgroup interaction p=0.6500

Treatment-by-time-by-subgroup interaction p=0.6487
Nintedanib slows disease progression irrespective of the DL\textsubscript{co} at baseline

To assess the impact of DL\textsubscript{co} % pred. at baseline on the treatment effect of nintedanib, Maher et al. analyzed patients with DL\textsubscript{co} >40% versus ≤40% pred. at baseline.\textsuperscript{28} The data suggest that the treatment effect of nintedanib is independent of DL\textsubscript{co} % pred. at baseline. However, in the group of patients with DL\textsubscript{co} ≤40% pred. at baseline (compared to >40% pred. at baseline):

- More patients experienced an acute IPF exacerbation (but no significant difference in the treatment effect of nintedanib between groups was detected)
- Patients reported a greater increase in SGRQ total score (equaling a worsening of quality of life) and these patients also displayed a more pronounced treatment effect with nintedanib

### Annual rate of decline in FVC by DL\textsubscript{co} % predicted at baseline

<table>
<thead>
<tr>
<th>DL\textsubscript{co} &gt;40% predicted</th>
<th>DL\textsubscript{co} ≤40% predicted</th>
</tr>
</thead>
<tbody>
<tr>
<td>Δ103.1 mL (95% CI: 63.6, 142.6)</td>
<td>Δ124.3 mL (95% CI: 56.2, 192.4)</td>
</tr>
<tr>
<td>(n=428)</td>
<td>(n=281)</td>
</tr>
<tr>
<td>Adjusted annual rate (SE) of decline in FVC (mL/year)</td>
<td>Adjusted annual rate (SE) of decline in FVC (mL/year)</td>
</tr>
<tr>
<td>-104.7</td>
<td>-136.4</td>
</tr>
<tr>
<td>-207.8</td>
<td>-260.7</td>
</tr>
</tbody>
</table>

- Treatment-by-time-by-subgroup interaction p=0.5492
FVC decline over 24 weeks can predict mortality, but not FVC decline in the following weeks

An analysis aimed to explore the impact of change in FVC in the first 24 weeks on subsequent FVC decline and mortality up to week 52 in patients with IPF. Patients were grouped according to the absolute FVC declines of <5%, ≥5%, or ≥10% pred. from baseline to week 24 and changes in FVC % predicted and mortality between weeks 24 and 52. The analysis showed that the FVC decline from baseline to week 24 did not predict FVC decline in the following 24 weeks. Irrespective of the change in FVC % pred. in the first 24 weeks, a greater proportion of patients treated with nintedanib had an increase/no decline in FVC % predicted between week 24 and week 52 compared to patients on placebo. Nevertheless, it could be demonstrated that an absolute FVC decline of ≥5% or ≥10% pred. in the first 24 weeks was associated with higher mortality in the following 24 weeks.

No significant effect of baseline statin use on the benefit of nintedanib

Since the potential effect of statins in combination with anti-fibrotic treatment is unknown, a post-hoc subgroup analysis of patients enrolled in the INPULSIS® trials and using statins at baseline was conducted. The analysis showed no significantly different rates of decline in FVC % pred. between the subgroups. However, the annual rate of decline in FVC was numerically lower in patients receiving statins at baseline and this result should be validated in prospective clinical trials.
Pirfenidone in IPF: RECAP\textsuperscript{21} subgroup analyses

Treatment with pirfenidone in patients with GAP1

The treatment effect of pirfenidone on patients with GAP1 was compared in a subgroup analysis of patients who were enrolled in CAPACITY004 and continued in RECAP (n=144).\textsuperscript{31}

The analysis compared patients who received pirfenidone 2403 mg/d in CAPACITY004 to those who started CAPACITY004 on placebo and changed to pirfenidone in RECAP. It shows maintained rate of reduction in decline of FVC % pred. over 180 weeks.

![Treatment with pirfenidone: Annual rate of decline (FVC\% predicted) with GAP1 (n=144)](image)

Treatment with pirfenidone in patients with severe lung function impairment (FVC % pred. <50%)

A subgroup analysis evaluated the benefit of pirfenidone in patients with a more severe impairment of lung function (FVC <50% pred.) at baseline in RECAP (n=54).\textsuperscript{32}

Compared to patients with higher FVC at baseline (n=530), the patients with FVC <50% pred. treated with pirfenidone showed a similar rate of decline and also a similar safety profile.

![Mean FVC % predicted at RECAP entry (n=584)](image)

![Annual rate of decline in RECAP (n=584)](image)
Pirfenidone in IPF: Pooled ASCEND/CAPACITY\textsuperscript{22,23} subgroup analyses

Continued treatment with pirfenidone after a decline in 6MWD $\geq 15\%$

Nathan et al. investigated the effect of continued treatment with pirfenidone after a decline in 6MWD $\geq 15\%$ within the first 6 months of treatment.\textsuperscript{33} Outcomes were analyzed 6 months after initial decline in 6MWD. The authors reported a continued treatment benefit with pirfenidone for patients who experienced a 6MWD decline $\geq 15\%$ compared to patients treated with placebo. Significant protective benefit was detected against further decline FVC and 6MWD (or death).

**Effect of pirfenidone by baseline characteristics and disease activity measures**

A post hoc analysis of the pooled data validated the significant benefit of treatment with pirfenidone on lung function across various subgroups.\textsuperscript{34} The subgroups were defined by demographics and baseline disease activity measures for 623 patients in the pirfenidone group and 624 in the placebo group. The annualized rate of FVC decline favored pirfenidone over placebo regardless of age, sex, lung function subgroups (DL\textsubscript{CO}, FVC, FEV\textsubscript{1}/FVC) supplemental oxygen or GAP stage.
Pirfenidone dose modifications and dose intensity

Investigation of dose modifications and treatment interruptions during the ASCEND\textsuperscript{22} and CAPACITY\textsuperscript{23} trials showed that dose adjustments occurred early (~95 days) and with limited duration (median dose reduction of 28 days, median dose interruption of 14 days).\textsuperscript{35}

Of patients treated with pirfenidone (n=623),
• 46% had to reduce dose, compared to 29% of patients treated with placebo (n=624)
• 41% had to interrupt treatment, compared to 25% of patients treated with placebo (n=624)

Dose intensity of >90% was observed in
• 424 out of 623 patients on pirfenidone
• 559 out of 624 patients on placebo

Mean difference (±SEM) in annual rate of FVC decline between groups over 52 weeks
• Dose intensity >90%: 105.5±18.4 mL (p<0.0001)
• Dose intensity ≤90%: 102.1±43.4 mL (p=0.0191)

In addition, analysis of data from CAPACITY 004 showed that patients on pirfenidone exhibit a dose-dependent change from baseline in FVC % predicted.

Predicting life expectancy and response to treatment with pirfenidone treatment

Fisher et al. compared the predicted life expectancies for patients with IPF treated with pirfenidone or best supportive care (BSC).\textsuperscript{36} For pirfenidone, the investigators used Kaplan-Meier survival data from CAPACITY, ASCEND and RECAP while data from two IPF registries were used for BSC. Despite this being a cross-trial comparison, the model suggests a significantly improved life expectancy by almost 3 years with pirfenidone compared with BSC in patients with IPF.
Clinical characteristics of patients with ILD and IPF in the real world

German registry reports many ILD patients with severe disease and with ILD-associated hospitalizations
The German EXCITING-ILD researchers presented several characteristics of the ILD patients enrolled in the registry (started in 10/2014; data cut-off 1/2016; n=201). Notably, they found that many patients presented with severe disease (measured by GAP-ILD index) and that ILD associated hospitalizations occur often (47% of patients included in the registry had been hospitalized within the 6 months before enrolling in the registry and of these, 65% were hospitalized for ILD reasons).

Incidence of IPF and CTD-ILD in India
For the first time, incidence of IPF and CTD-ILD in India was investigated in the ILD-India registry: 1084 Indian patients with newly onset ILD were evaluated by MDD among ILD experts and key characteristics for those patients newly diagnosed with IPF (per 2011 criteria) and CTD-ILD were reported.

Quality of life of patients with IPF affected by lung function and comorbidities
A group of investigators around Dr. Kreuter found a close relationship between lung function, comorbidities and quality of life, measured by 3 scores (EQ-5D-VAS, EQ-5D index and SGRQ).• The quality of life of 572 patients with IPF from the INSIGHTS-IPF registry worsened significantly for all scores with increasing number of comorbidities (p<0.001)
• FVC declines >10% showed significant negative effects for all scores
• A change of over 6% of FVC predicted was associated with a change of SGRQ total score of over 4 points, which is deemed clinically relevant

Smoking status in patients with ILD and IPF
A lot of patients with ILD in general, and with IPF in particular, have a history of smoking. At ERS, several groups reported on the smoking status of patients included in their registries:
• 57% of patients with ILD enrolled in the German EXCITING ILD registry (n=201) are current or ex-smokers.
• 44% of newly diagnosed patients with IPF in India were smokers (ILD-INDIA, n=148)
• 63% of British patients with IPF (BTS-IPF registry, n=767) were current or former smokers.
Predicting disease progression and mortality in IPF

Progression free survival in Australian patients with IPF below and above 80% FVC predicted

Patients with IPF from the Australian IPF registry (n=631) were analyzed with regards to progression free survival (PFS) above and below 80% FVC predicted.\textsuperscript{39} PFS was defined as decline in FVC>10% or DL\textsubscript{CO}>15% or death).

Factors associated with PFS:
- Male gender
- Impaired quality of life (SGRQ)
- Depression
- Cough severity
- Lower baseline FVC and DL\textsubscript{CO}

Characteristics of patients with FVC>80% predicted (n=235):
- Older age (p=0.001)
- Female sex (p<0.001)
- Improved PFS (HR 1.61; 95% CI 1.3,2.0; p<0.001)

However, 18% (n=41) of those patients still progressed at 12 months.

\textbf{DL\textsubscript{CO} predicts mortality better than FVC}

In order to find the best indicator of predicting mortality in IPF, investigators compared the accuracy of GAP and CPI scores as well as several univariate parameters in patients with IPF (n=209).\textsuperscript{40}

While DL\textsubscript{CO} was the only significant parameter in all multivariate models (p=<0.001), all three lung function parameters (DL\textsubscript{CO}, FVC and FEV\textsubscript{1}) were found significant for predicting mortality alone, while age and gender were not. The analysis revealed that DL\textsubscript{CO} predicts mortality better than FVC or FEV\textsubscript{1}.

\textbf{Extent of emphysema in patients with IPF has an impact on FVC decline}

In an analysis of 455 patients with IPF, patients with over 15% emphysema on HRCT showed less decline in FVC over 48 weeks than those without or with emphysema below 15%.\textsuperscript{41}

\textbf{Survival and lung function parameters not influenced by HRCT pattern}

A subgroup analysis of patients from the Czech population of the EMPIRE\textsuperscript{1} registry (n=513) showed that survival (from diagnosis up to 84 months) and lung function parameters are not influenced by the HRCT pattern of the patient at diagnosis.\textsuperscript{42} The investigators compared patients previously diagnosed with IPF who, on HRCT, showed UIP pattern (n=423), possible UIP pattern (n=71) or a pattern inconsistent with UIP (n=19).

There were no significant differences in survival (Kaplan-Meier survival curves and median survival) or in lung function (FVC and DL\textsubscript{CO} values at the time of diagnosis or rate of decline) between the groups, indicating that patients with atypical HRCT patterns should be treated like patients with UIP pattern.
ERS 2016 Congress Highlights – Interstitial Lung Disease (ILD)
London, UK | September 3rd – 7th 2016


Glossary

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<th>Abbreviation</th>
<th>Definition</th>
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<tr>
<td>6MWD</td>
<td>6-minute walk distance</td>
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<tr>
<td>AAT</td>
<td>Antiacid therapy</td>
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<td>ADR</td>
<td>Adverse drug reaction</td>
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<td>AE</td>
<td>Adverse event</td>
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<tr>
<td>BSC</td>
<td>Best supportive care</td>
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<td>BTS</td>
<td>British Thoracic Society</td>
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<tr>
<td>CI</td>
<td>Confidence interval</td>
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<tr>
<td>CPI</td>
<td>Composite physiologic index</td>
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<td>CPU</td>
<td>Compassionate use program</td>
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<td>CTD-ILD</td>
<td>Connective tissue disease associated ILD</td>
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<td>DLco</td>
<td>Diffuse capacity of the lung for carbon monoxide</td>
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<td>EQ-5D</td>
<td>EuroQuol</td>
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<tr>
<td>EQ-5D-VAS</td>
<td>EuroQuol visual analogue scale</td>
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<td>ERS</td>
<td>European Respiratory society</td>
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<tr>
<td>FVC</td>
<td>Forced vital capacity</td>
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<td>GAP</td>
<td>Gender-Age-Physiology index</td>
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<td>GERD</td>
<td>Gastroesophageal reflux disease</td>
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<td>HR</td>
<td>Hazard ratio</td>
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<td>HRCT</td>
<td>High resolution computed tomography</td>
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<tr>
<td>HRQoL</td>
<td>Health-related quality of life</td>
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<tr>
<td>ILD</td>
<td>Interstitial lung disease</td>
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<tr>
<td>IPF</td>
<td>Idiopathic pulmonary fibrosis</td>
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<tr>
<td>MDD</td>
<td>Multi-disciplinary discussion</td>
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<td>MDT</td>
<td>Multi-disciplinary team</td>
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<tr>
<td>NAC</td>
<td>N-acetylcysteine</td>
</tr>
<tr>
<td>NPU</td>
<td>Named patient use</td>
</tr>
<tr>
<td>PFS</td>
<td>Progression free survival</td>
</tr>
<tr>
<td>SAE</td>
<td>Serious adverse event</td>
</tr>
<tr>
<td>SGRO</td>
<td>St George’s Respiratory Questionnaire</td>
</tr>
<tr>
<td>SLB</td>
<td>Surgical lung biopsy</td>
</tr>
<tr>
<td>TEAE</td>
<td>Treatment emergent AE</td>
</tr>
<tr>
<td>UIP</td>
<td>Usual Interstitial Pneumonia</td>
</tr>
<tr>
<td>VC</td>
<td>Vital Capacity</td>
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</table>

Explore the educational resources in the IPF learning center and find out more about nintedanib’s safety and efficacy on global.OFEV.com!