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Survival and course of lung function in the presence or absence of antifibrotic treatment in patients with idiopathic pulmonary fibrosis: long-term results of the INSIGHTS-IPF registry

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Survival was significantly higher in antifibrotic-treated (AT) IPF patients, but the course of lung function parameters was similar in AT and non-AT patients, suggesting that functional stability alone may not safeguard against premature mortality in IPF https://bit.ly/2RDsrVY

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ABSTRACT

Objective: There is a paucity of observational data on antifibrotic therapy for idiopathic pulmonary fibrosis (IPF). We aimed to assess the course of disease of IPF patients with and without antifibrotic therapy under real-life conditions.

Methods: We analysed data from a non-interventional, prospective cohort study of consecutively enrolled IPF patients from 20 interstitial lung disease expert centres in Germany. Data quality was ensured by automated plausibility checks, on-site monitoring, and source data verification. Propensity scores were applied to account for known differences in baseline characteristics between patients with and without antifibrotic therapy.

Results: Among the 588 patients suitable for analysis, the mean±SD age was 69.8±9.1 years, and 81.0% were male. The mean±SD duration of disease since diagnosis was 1.8±3.4 years. The mean±SD value at baseline for forced vital capacity (FVC) and diffusion capacity (D_{LCO}) were 68.6±18.8% predicted and 37.8±18.5% predicted, respectively. During a mean±SD follow-up of 1.2±0.7 years, 194 (33.0%) patients died. The 1-year and 2-year survival rates were 87% *versus* 46% and 62% *versus* 21%, respectively, for patients with *versus* without antifibrotic therapy. The risk of death was 37% lower in patients with antifibrotic therapy (hazard ratio 0.63, 95% CI 0.45; 0.87; p=0.005). The results were robust (and remained statistically significant) on multivariable analysis. Overall decline of FVC and D_{LCO} was slow and did not differ significantly between patients with or without antifibrotic therapy.

Conclusions: Survival was significantly higher in IPF patients with antifibrotic therapy, but the course of lung function parameters was similar in patients with and without antifibrotic therapy. This suggests that in clinical practice, premature mortality of IPF patients eventually occurs despite stable measurements for FVC and D_{LCO}.