ORIGINAL ARTICLE
INTERSTITIAL LUNG DISEASE

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The natural history of progressive fibrosing interstitial lung diseases

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Analyses of data from the INBUILD and INPULSIS trials suggest that progressive fibrosing ILDs other than IPF have a clinical course similar to IPF, irrespective of underlying ILD diagnosis or the fibrotic pattern on HRCT http://bit.ly/3apG0Q5

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ABSTRACT We used data from the INBUILD and INPULSIS trials to investigate the natural history of progressive fibrosing interstitial lung diseases (ILDs).

Subjects in the two INPULSIS trials had a clinical diagnosis of idiopathic pulmonary fibrosis (IPF) while subjects in the INBUILD trial had a progressive fibrosing ILD other than IPF and met protocol-defined criteria for ILD progression despite management. Using data from the placebo groups, we compared the rate of decline in forced vital capacity (FVC) (mL·year^{-1}) and mortality over 52 weeks in the INBUILD trial with pooled data from the INPULSIS trials.

The adjusted mean annual rate of decline in FVC in the INBUILD trial (n=331) was similar to that observed in the INPULSIS trials (n=423) (−192.9 mL·year^{-1} and −221.0 mL·year^{-1}, respectively; nominal p-value=0.19). The proportion of subjects who had a relative decline in FVC >10% predicted at Week 52 was 48.9% in the INBUILD trial and 48.7% in the INPULSIS trials, and the proportion who died over 52 weeks was 5.1% in the INBUILD trial and 7.8% in the INPULSIS trials. A relative decline in FVC >10% predicted was associated with an increased risk of death in the INBUILD trial (hazard ratio 3.64) and the INPULSIS trials (hazard ratio 3.95).

These findings indicate that patients with fibrosing ILDs other than IPF, who are progressing despite management, have a subsequent clinical course similar to patients with untreated IPF, with a high risk of further ILD progression and early mortality.