Progressive fibrosing interstitial lung disease: we know it behaves badly, but what does that mean?

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The criteria used to define progressive fibrosing interstitial lung disease identify a rapidly progressive phenotype that behaves like idiopathic pulmonary fibrosis, regardless of the underlying diagnosis.

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The concept behind the INBUILD clinical trial was simple: if nintedanib slows progression of idiopathic pulmonary fibrosis (IPF) [1], then it might also slow progression in other forms of fibrotic interstitial lung disease (ILD) [2]. This was recently shown to be the case [3], with nintedanib slowing the rate of ILD progression in a diverse cohort of patients without IPF who met specific criteria for recent worsening. This study was a major advance, identifying a new treatment for a group of patients who previously had limited options.