Progressive pulmonary fibrosis: an expert group consensus statement

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Shareable abstract (@ERSpublications)
Progressive pulmonary fibrosis (PPF) explains what clinicians increasingly face in practice. Assessing ILD progression, its risk and improved treatments based on current evidence for PPF (despite initial management) form the mainstay of this document. http://bit.ly/3GLdqfs

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Abstract

This expert group consensus statement emphasises the need for standardising the definition of progressive fibrosing interstitial lung diseases (F-ILDs), with an accurate initial diagnosis being of paramount importance in ensuring appropriate initial management. Equally, case-by-case decisions on monitoring and management are essential, given the varying presentations of F-ILDs and the varying rates of progression. The value of diagnostic tests in risk stratification at presentation and, separately, the importance of a logical monitoring strategy, tailored to manage the risk of progression, are also stressed. The term "progressive pulmonary fibrosis" (PPF) exactly describes the entity that clinicians often face in practice. The importance of using antifibrotic therapy early in PPF (once initial management has failed to prevent progression) is increasingly supported by evidence. Artificial intelligence software for high-resolution computed tomography analysis, although an exciting tool for the future, awaits validation. Guidance is provided on pulmonary rehabilitation, oxygen and the use of non-invasive ventilation focused specifically on the needs of ILD patients with progressive disease. PPF should be differentiated from acute deterioration due to drug-induced lung toxicity or other forms of acute exacerbations. Referral criteria for a lung transplant are discussed and applied to patient needs in severe diseases where transplantation is not realistic, either due to access limitations or transplantation contraindications. In conclusion, expert group consensus guidance is provided on the diagnosis, treatment and monitoring of F-ILDs with specific focus on the recognition of PPF and the management of pulmonary fibrosis progressing despite initial management.