

PROGRESSIVE PULMONARY FIBROSIS IN LIGHT OF THE ATS/ERS/ JRS/ALAT 2022 CLINICAL GUIDELINES

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Abstract

Idiopathic pulmonary fibrosis – is a specific form of chronic, progressive, fibrosing interstitial pneumonia of unknown cause, which occurs primarily in 50 years and older patients, limited to the lungs, and is associated with the histopathologic and/or radiologic pattern of usual interstitial pneumonia (UIP).

In 2000 American thoracic society (ATS) and European respiratory society (ERS) published the first international; statement on diagnosis and treatment of IPF American Thoracic Society, European Respiratory Society. Idiopathic pulmonary fibrosis: diagnosis and treatment. International consensus statement. In 2011 there was published a new statement on diagnosis and treatment of IPF, approved by Japan respiratory society (JTS) and Latin American thoracic society (ALTS) – An Official ATS/ERS/JRS/AL : Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. In 2015 section “Treatment” and in 2018 section “Diagnosis” were revised.

It is known that in part of patients with such interstitial lung diseases (ILD) as idiopathic interstitial pneumonia, systemic sclerosis, pneumoconiosis, chronic hypersensitivity pneumonitis, sarcoidosis the disease may acquire a progressive uncontrolled development with the combination of symptoms called progressive pulmonary fibrosis (PPF). At the same time the results of successful use of antifibrotic therapy have been published recently (SENSCIS, INBUILD studies).

All this provided the rationale for extended indications for use of antifibrotic therapy to cover other ILDs with the features of PPF. In this connection, ATS, ERS, JRS and ALAT experts published in May 2022 An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults, in which along with the partial changes regarding the principles of diagnosis and treatment of IPF, the definition, diagnosis criteria and management of PPF due to other fibrosing ILDs were presented.

The article presents the major statements of new guideline regarding the terminology, diagnosis and treatment of PPF.

Key words: progressive pulmonary fibrosis, definition, diagnosis criteria, treatment.

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