IDIOPATHIC PULMONARY FIBROSIS AND PROGRESSIVE PULMONARY FIBROSIS IN ADULTS: ADAPTED EVIDENCE-BASED CLINICAL GUIDELINE (DRAFT)

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Abstract

In 2000 American Thoracic Society (ATS) and European Respiratory Society (ERS) published first international statement on diagnosis and treatment of idiopathic pulmonary fibrosis (IPF) — American Thoracic Society, European Respiratory Society. Idiopathic pulmonary fibrosis: diagnosis and treatment. International consensus statement. In 2011 there was published new statement on diagnosis and treatment of IPF, approved by ATS, ERS, Japan Respiratory Society (JRS) and Latin American Thoracic Society (ALAT) — 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» an update.

It is known, that in part of the patients with such an insterstitial lung disease (ILD) as idiopathic nonspecific interstitial pneumonia, systemic sclerosis, pneumoconiosis, chronic hypersensitivity pneumonitis, sarcoidosis, lung fibrosis may acquire a progressive uncontrolled character with combination of symptoms described as Progressive Pulmonary Fibrosis (PPF). Besides, in recent years there have been published data on successful use of antifibrotic therapy in several fibrosing ILD, other than IPF (INBUILD, SENSCIS studies). This has required the change of the treatment paradigm in favor for a unified approach to antifibrotic therapy.

This was a rationale for an inclusion of other ILD manifested as PPF into the list of indications for use of antifibrotic therapy. To comply with this novelty the experts of ATS, ERS, JRS and ALAT in May 2022 published new statement An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults. In this document alongside with partial update of IPF diagnosis and treatment principles, there have been presented a definition, diagnosis criteria and recommendations for treatment of PPF in other ILD.

In summary, «An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management» (2011) and its updated sections «An Official ATS/ERS/JRS / ALAT Clinical Practice Guideline: Treatment of Idiopathic Pulmonary Fibrosis. An Update of the 2011 Clinical Practice Guideline» (2015), "Diagnosis of Idiopathic Pulmonary Fibrosis: An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline» (2018) and Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults were

used a the prototype documents for creation of Adapted clinical guideline «Idiopathic pulmonary fibrosis and progressive pulmonary fibrosis in adults».

Key words: idiopathic pulmonary fibrosis, progressive pulmonary fibrosis, definiition, diagnosis, treatment, antifibrotic therapy.

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