EVOLUTION OF PRINCIPLES OF DIAGNOSIS AND TREATMENT OF IDIOPATHIC PULMONARY FIBROSIS IN THE INTERNATIONAL GUIDELINE STATEMENTS

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Abstract

Idiopathic pulmonary fibrosis (IPF) is specific form of chronic progressive interstitial fibrosing pneumonia of unknown nature, mainly occurring in patients > 50 years of age, limited to the lungs and associated with histological and/or radiological pattern of usual interstitial pneumonia.

Epidemiological studies estimate that prevalence of ILF in different countries varies between 1,25 and 63 cases per 100 000 persons. Along with that, ILF is characterized by unfavorable prognosis — median survival time ranges within 2,5–3,5 years from the time of diagnosis.

In 2000 American thoracic society (ATS) and European respiratory society (ERS) published first international statement on diagnosis and treatment of ILF — American Thoracic Society, European Respiratory Society. Idiopathic pulmonary fibrosis: diagnosis and treatment. International consensus statement.

Data from studies, accumulated during next 10 years of research, determined the necessity of update of certain diagnostic criteria and principles of therapy. In this regard, a new guideline for diagnosis and treatment of IPF was published in 2011 and approved by ATS, ERS, Japanese Respiratory Society (JRS) and Latin American Thoracic Association (ALAT) — An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. A new update on "Treatment of IPF" was published in 2015, and chapter "Diagnosis" was updated later in 2018.

Current literature review focuses on the principles and algorithms of IPF treatment and the changes in guidelines, occurred from the time of first Statement published.

Summarizing review results, we can conclude, that evolution of diagnostics principles, which limited the indications for surgical lung biopsy for the purpose of diagnosis verification, is caused by explosive technological advances in the field of chest computed tomography. The capability of computed tomography has grown to such an extent, that in terms of morphological diagnosing this method can compete with histological examination.

The changes in management principles were evoked by the revision of IPF pathogenesis mechanisms. Since 2000, most of the experts concluded, that fibrosis, rather than inflammation, was the leading link of pathogenesis. A fibrosing process, initially playing a reparative role, further due to unknown reasons, gains an uncontrolled progressive character. Accordingly, antifibrotic compounds, such as pirfenidone and nintedanib, came to substitute glucocorticosteroids, known for their powerful anti-inflammatory potential.

The publication also presents the review of most relevant randomized clinical trials on safety and efficacy of antifibrotic drugs — pirfenidone and nintedanib.

Key words: idiopathic pulmonary fibrosis, diagnostics, treatment, algorithms, pirfenidone, nintedanib.

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