## PRINCIPLES OF TREATMENT OF IDIOPATHIC PULMONARY FIBROSIS PATIENTS

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Abstract

Increase of effectiveness of treatment of idiopathic pulmonary fibrosis (IPF) patients in one of the most complicated problems in respiratory care at present.

In the last 15 years the principles of therapy of IPF patients have changed substantially, which happened due to evolution of the disease pathogenesis understanding.

The recommendations of first guidelines on diagnosis and management of IPF (2000) were based on the concept of chronic interstitial disease, affecting mainly alveoli (alveolitis) with fibrosis as an end-stage outcome of inflammation. Considering this an aggressive anti-inflammatory therapy was recommended. The glucocorticosteroids (GCS) and cytotoxic agents (CA) were the cornerstone of this therapy.

A new paradigm of IPF pathogenesis was introduced in 2001, which denied the leading role of inflammation in pathogenesis of IPF. A new concept is based on a hypothesis of alveolar epithelial cells injury by an unidentified factor at the presence of inadequate reparative response. Despite a certain significance of inflammatory mediators in the development and progression of IPF, the concept of advanced lung fibrosis as a consequence of alveolar epithelium damage has strong scientific grounds. Taking that, the priority district of the research in IPF management is a development of new pharmacological compounds, exhibiting anti-fibrotic effects.

The article presents characteristics of new anti-fibrotic medicines — pirfenidone and nintedanibe, recommended for the management of IPF in 2015 edition of ATS/ERS/JRS/ALAT Guidelines. At the same time the feasibility and possible benefits of GCS and CA use in some IPF cases is under discussion.

**Key words**: idiopathic pulmonary fibrosis, treatment, anti-fibrosis medicines.

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