AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

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Abstract

Background: This American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax guideline updates prior idiopathic pulmonary fibrosis (IPF) guidelines and addresses the progression of pulmonary fibrosis in patients with interstitial lung diseases (ILDs) other than IPF.

Methods: A committee was composed of multidisciplinary experts in ILD, methodologists, and patient representatives. *1*) Update of IPF: Radiological and histopathological criteria for IPF were updated by consensus. Questions about transbronchial lung cryobiopsy, genomic classifier testing, antacid medication, and antireflux surgery were informed by systematic reviews and answered with evidence-based recommendations using the Grading of Recommendations, Assessment, Development and Evaluation (GRADE) approach. *2*) Progressive pulmonary fibrosis (PPF): PPF was defined, and then radiological and physiological criteria for PPF were determined by consensus. Questions about pirfenidone and nintedanib were informed by systematic reviews and answered with evidence-based recommendations using the GRADE approach.

Results: *1*) Update of IPF: A conditional recommendation was made to regard transbronchial lung cryobiopsy as an acceptable alternative to surgical lung biopsy in centers with appropriate expertise. No recommendation was made for or against genomic classifier testing. Conditional recommendations were made against antacid medication and antireflux surgery for the treatment of IPF. *2*) PPF: PPF was defined as at least two of three criteria (worsening symptoms, radiological progression, and physiological progression) occurring within the past year with no alternative explanation in a patient with an ILD other than IPF. A conditional recommendation was made for nintedanib, and additional research into pirfenidone was recommended.

Conclusions: The conditional recommendations in this guideline are intended to provide the basis for rational, informed decisions by clinicians.

Keywords: idiopathic pulmonary fibrosis; progressive pulmonary fibrosis; radiology; histopathology

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