New guideline aids in diagnosing idiopathic pulmonary fibrosis

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A new international guideline has been developed to help physicians diagnosis idiopathic pulmonary fibrosis (IPF), a rare and often fatal lung disease whose cause is unknown.

The 2018 clinical practice guideline was developed by experts representing four major respiratory societies—the American Thoracic Society (ATS), the European Respiratory Society (ERS), Japanese Respiratory Society (JRS) and the Latin American Thoracic Society (ALAT)—and published online and in the American Thoracic Society's Sept. 1 edition of the *American Journal of Respiratory and Critical Care Medicine*.

IPF is the most common and deadly form of a group of more than 200 conditions known broadly as interstitial lung disease. Most often, IPF is diagnosed in adults over age 60 and more often in men than women. Symptoms include shortness of breath, a dry cough and low oxygen levels. The median survival is three to five years after the diagnosis.

"Diagnosing IPF is challenging because these symptoms are non-specific: they occur with all other interstitial lung diseases and with other respiratory problems," said Ganesh Raghu, MD, chair of the guideline committee and professor of medicine and director of the Center for Interstitial Lung Disease at the University of Washington in Seattle. "Because drugs may slow the progression of IPF, an early and accurate diagnosis is essential for prompt and appropriate treatment for this fatal disease."

The 29-member guideline committee included international clinical and scientific experts and an IPF patient. The committee discussed the findings of all accumulated evidence pertinent to IPF, rated the strength of those findings using the GRADE (Grading of Recommendations, Assessment, Development and Evaluation) system and made recommendations.

"The approach used to develop the guidelines adhered strictly to the state-of-the-art Institute of Medicine Standards for Trustworthy Guidelines," said Kevin C. Wilson, MD, lead methodologist and project manager, as well as professor of medicine at Boston University School of Medicine. "This should provide the health care community with confidence that the recommendations are as unbiased and evidence-based as possible."

The 2018 guideline is an update of IPF diagnostic guidelines the four respiratory societies produced in 2011.

"The 2011 guideline provided the first evidence-based, formal criteria for diagnosis of IPF and allowed patients with a well-defined diagnosis of IPF to participate in numerous clinical studies and randomized controlled trials that enhanced our understanding of the disease," Dr. Raghu said. "However, it became clear that there were significant challenges in ascertaining the diagnosis per the 2011 criteria, and abundant evidence accumulated since then allowed the committee to refine the diagnostic criteria now."

Chief among the refinements included in the criteria is the use of four diagnostic categories based on high-resolution tomography (HRCT) of the lung: usual interstitial pneumonia (UIP) pattern, probable UIP pattern, indeterminate pattern and alternative diagnosis.

The committee recommended the same terminology for both images produced using HRCT of the lung and microscopic analysis of biopsied

tissue. Consistent categorization should facilitate multidisciplinary discussions (MDD) between a pulmonologist (and in some cases, a rheumatologist), radiologist and pathologist of a diagnosis, the committee wrote.

The committee made the following recommendations for diagnosing IPF in all adult patients with newly detected interstitial lung disease (ILD) of unknown cause:

- For all patients, a detailed history should be taken of both medication use and environmental exposures and serological testing should be performed to exclude connective tissue disease as a potential cause of the ILD.
- For patients with a HRCT pattern of probable UIP, indeterminate, or an alternative diagnosis, conditional recommendations were made for performing bronchoalveolar lavage (BAL) and surgical lung biopsy (SLB); due to lack of evidence, no recommendation was made for or against performing transbronchial lung biopsy (TBBx) or lung cryobiopsy.
- Conditional recommendation for MDD to aid in diagnosing IPF, particularly when the HRCT pattern has features of probable UIP pattern, indeterminate or alternative diagnosis.
- In contrast, for patients with newly detected ILD who have a HRCT pattern of UIP, strong recommendations were made against performing SLB, TBBx and lung cryobiopsy; a conditional recommendation was made against performing BAL.
- Strong recommendation against measurement of serum biomarkers for the sole purpose of distinguishing IPF from other ILDs.

"Our hope is that this new guideline will bridge the gap between the experienced IPF experts and general pulmonologists in making a prompt

and accurate diagnosis of IPF for the individual unfortunately confronted with the disease," Dr. Raghu said. "This will allow patients to make well-informed decisions about treatment options and participation in clinical trials."

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