Editorial

The new ATS-ERS-JRS-ALAT
Guidelines for diagnosis of Idiopathic Pulmonary Fibrosis (IPF)

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Idiopathic Pulmonary Fibrosis (IPF) is the most frequent idiopathic interstitial "pneumonia"[1-3] and one of the almost 500 interstitial lung diseases. IPF is a specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause occurring in adults. Radiologic and/or histopathologic patterns are consistent with usual interstitial pneumonia (UIP)[4-6].

The diagnosis of IPF is a difficult and dynamic one. According to existing guidelines[1] it is based on the exclusion of known causes of interstitial lung disease and the presence of a UIP pattern on high resolution computed tomography (HRCT) or the presence of a definite or possible UIP pattern on HRCT with a surgical lung biopsy showing definite or possible UIP pattern. In this way a number of combinations arise (Table 1) making the diagnosis questionable in certain cases, making necessary the reference of the case to a specialized center where a multidisciplinary discussion (MDD) will decide if the data are enough to diagnose IPF or the case remain unclassifiable[5-7]. Under these circumstances the diagnosis in many cases can remain unclassifiable and clarification of diagnostic interventions as defined in the 2011 guidelines is the subject of the ongoing development of new diagnostic guidelines under the auspices of the American Thoracic Society, the European

TABLE 1. Table showing the various diagnostic combinations according to radiologic (HRCT) and histologic pattern. A number of combinations need further evaluation for their diagnostic accuracy.

Diagnosis of IPF by Lung Biopsy

Radiologic Pattern \ Histopathologic Pattern	UIP	Probable UIP	Possible UIP	Not UIP	Not performed
UIP	IPF	IPF	IPF	Not IPF	IPF
Possible UIP	IPF	IPF	+/- IPF	Not IPF	Not IPF
Inconsistent with UIP	+/- IPF	Not IPF	Not IPF	Not IPF	Not IPF

Raghu G, et al. *Am J Respir Crit Care Med.* 2011;183:788-824.

Respiratory Society, the Japan Respiratory society and the Latin American Thoracic Society (ATS/ERS/JRS/ALAT).

The chair of the committee is Ganesh Raghu (USA) and co-chairs Martine Remy-Jardine (EU), Jeff Myers (USA) and Luca Richeldi (EU). Pulmonologists are 18, radiologists 5 and pathologists 4 (Table 2). A number of specific questions are to be addressed utilizing full guideline methodology including PICO questions, systematic reviews, and the GRADE approach including but not limited to:

1. Genetic testing
2. Specific biomarkers
3. Volumetric HRCT.

TABLE 2. Members of the new diagnostic guidelines committee.

CHAIR: ***Ganesh Raghu*** (USA)
CO-CHAIRS: ***Martine Remy-Jardine*** (EU); ***Jeff Myers*** (USA); ***Luca Richeldi*** (EU)
METHODOLOGIST/ PROJECT MANAGER: ***Kevin Wilson*** (USA)
PROJECT COORDINATOR: ***Kimberly Lawrence*** (USA)
Members:

NORTH AMERICA (ATS):		
Name	**Area of Expertise**	**Location**
Jeff Myers	Pathologist	Ann Arbor, Michigan, USA
Fernando Martinez	Pulmonologist	New York, NY, USA
Harold Collard	Pulmonologist	San Francisco, CA, USA
David Lederer	Pulmonologist	New York, NY, USA
Sonye Danoff	Pulmonologist	Bethesda, Maryland, USA
Sudhakar Pipavath	Radiologist	Seattle, Washington DC, USA
Kevin Brown	Pulmonologist	Denver, Colorado, USA
Ella Kazerooni	Radiologist	Ann Arbor, Michigan, USA
William Travis	Pathologist	New York, NY, USA
Kevin Flaherty	Pulmonologist	Ann Arbor, Michigan, USA
Chris Ryerson	Pulmonologist	Vancouver, BC, CANADA
EUROPE (ERS):		
Martine Remy-Jardine	Radiologist	Lylle, FRANCE
Luca Richeldi	Pulmonologist	Rome, ITALY
Simon Walsh	Radiologist	London, UK
Andrew Nicholson	Pathologist	London, UK
Athol Wells	Pulmonologist	London, UK
Gisli Jenkins	Pulmonologist	Nottingham, UK
Juergen Behr	Pulmonologist	Munich, GERMANY
Vincent Cottin	Pulmonologist	Paris, FRANCE
Ferran Morell	Pulmonologist	Barcelona, SPAIN
Demosthenes Bouros	Pulmonologist	Athens, GREECE
MEXICO:		
Moises Selman	Pulmonologist/expertise in genetic markers	
JAPAN:		
Takeshi Johkow	Radiologist	
Yoshikazu Inoue-Gichi	Pulmonologist	
Azuma Arata	Pulmonologist	
Masanori Kitaichi	Pathologist	

Furthermore, interesting questions to be discussed are the following:

Should patients with newly detected ILD who are clinically suspected of having IPF and have a HRCT scan pattern consistent with probable or possible UIP undergo?

1. Transbronchial biopsy
2. Bronchoalveolar lavage
3. Surgical lung biopsy
4. Surgical lung biopsy more than one wedge lung biopsy from different parts of the same lung.
5. Multidisciplinary decision
6. Lung cryobiopsy
7. Lung tissue analyzed by molecular techniques.

Other significant questions for decisions are the following:

1. Should patients with newly detected ILD who are clinically suspected of having IPF but have the combination of: a) unclassifiable histopathology and b) a HRCT pattern of possible UIP or inconsistent with UIP be diagnosed with IPF?
2. Should patients with newly detected ILD who are clinically suspected of having IPF but have honeycomb cysts in the upper lobe on HRCT without air trapping be diagnosed with IPF?
3. Should we abandon the term 'idiopathic' as a prefix for pulmonary fibrosis?
4. In the absence of any clinical features of connective tissue disease how useful is serology especially in the elderly population?
5. How does the presence or absence of mutations affect our interaction with the patient and more importantly, should the recommendation be in favor of testing, how do we advise relatives about a 'positive' result – should they be tested and if so when and what should they do in the future?

These much needed updated guidelines are expected to elucidate many unresolved aspects of this devastating disease.

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