ITALIAN RESPIRATORY SOCIETY
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SOCIETÀ ITALIANA DI PNEUMOLOGIA

I T S
ITALIAN THORACIC SOCIETY

A I P O
ASSOCIAZIONE ITALIANA PNEUMOLOGI OSPEDALIERI

Convegno di presentazione della versione italiana delle linee guida per la diagnosi di fibrosi polmonare idiopatica

1 Febbraio 2020

Hotel Enterprise Milano

Linee guida ufficiali ATS/ ERS/JRS/ALAT per la pratica clinica

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L'ALGORITMO DIAGNOSTICO

Donato LACEDONIA

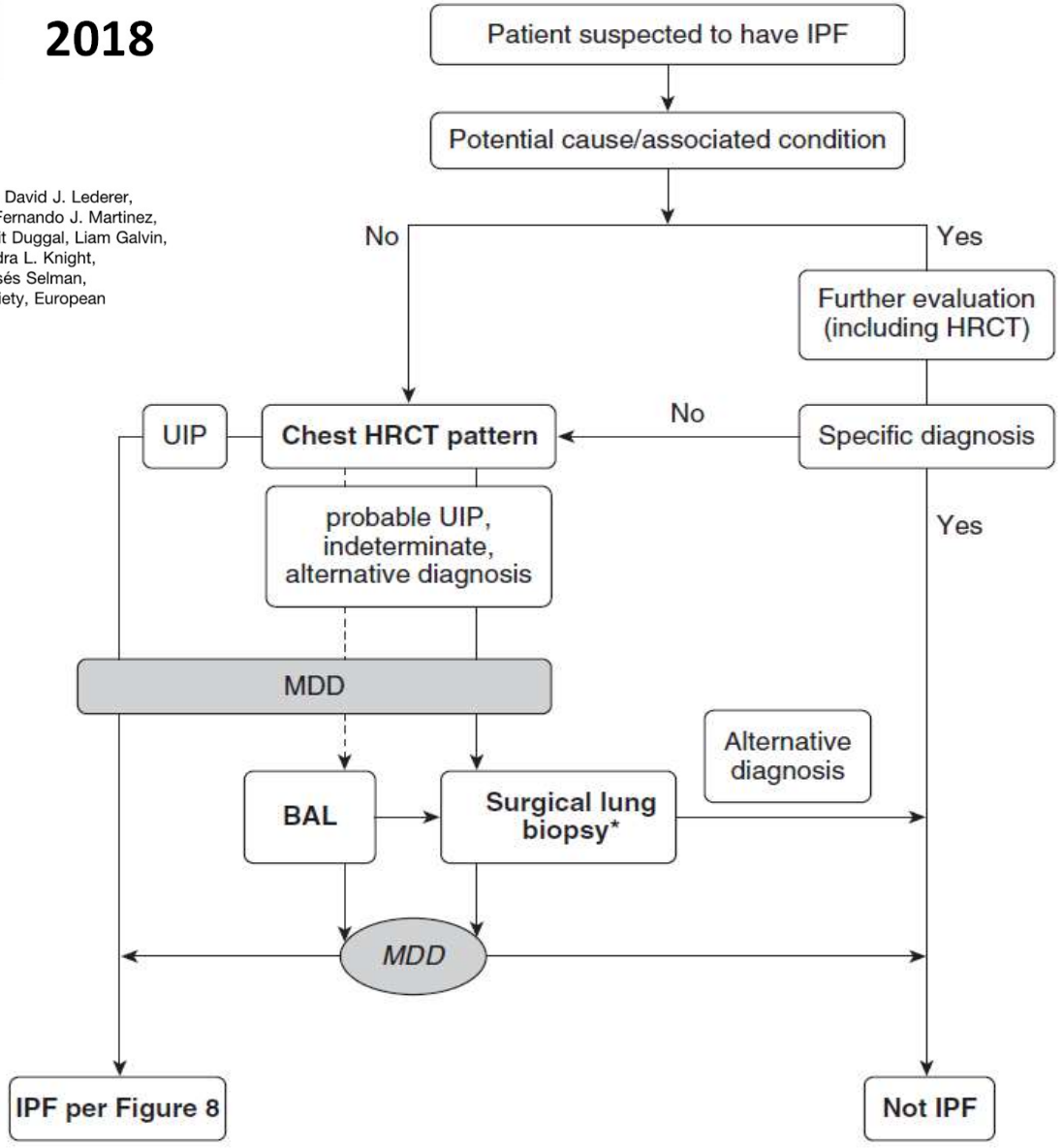
Università di Foggia

AMERICAN THORACIC SOCIETY DOCUMENTS **2018**

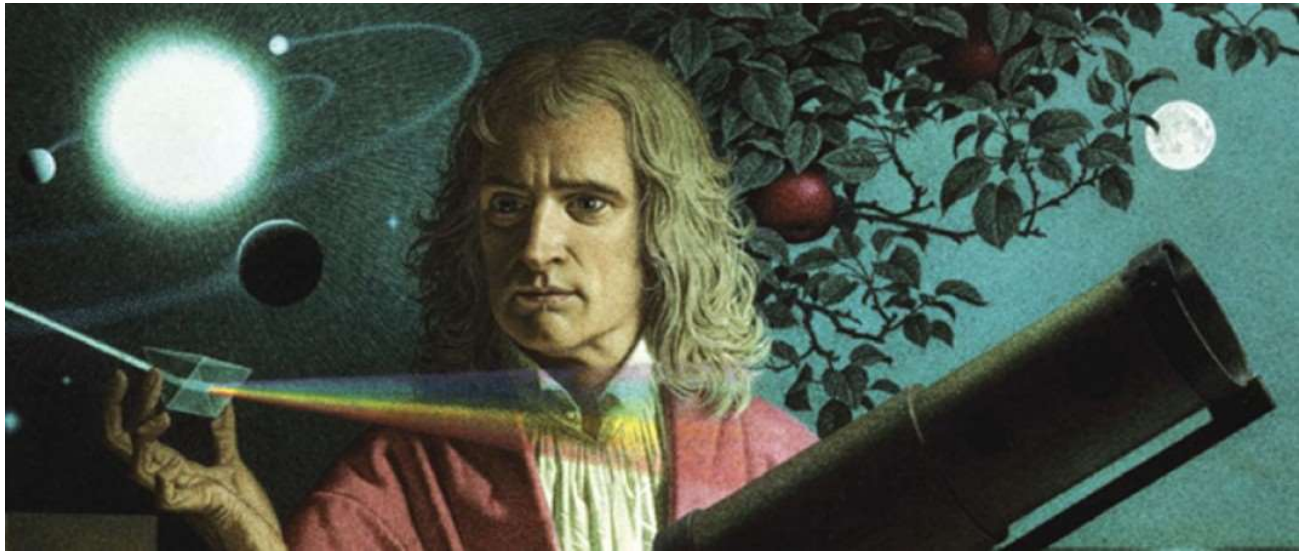
Diagnosis of Idiopathic Pulmonary Fibrosis

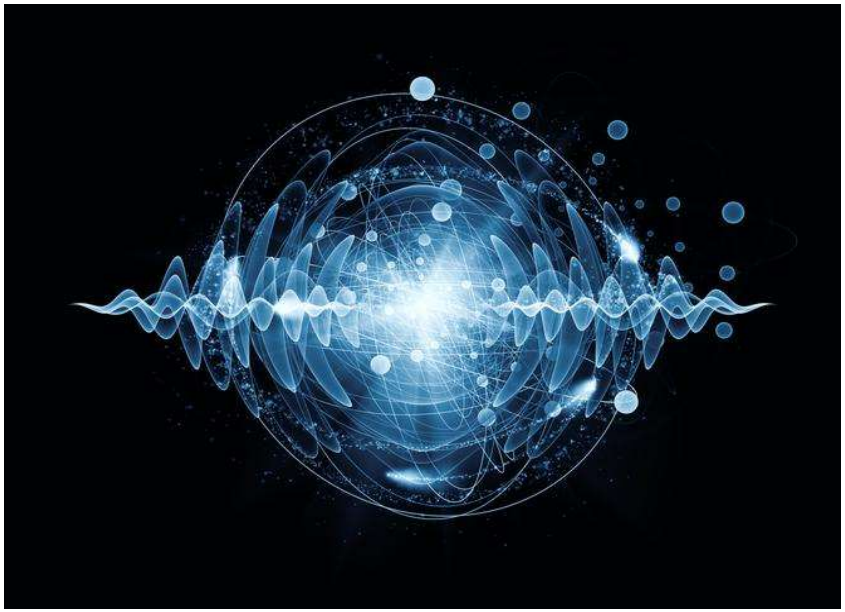
An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Jeffrey L. Myers, Luca Richeldi, Christopher J. Ryerson, David J. Lederer, Juergen Behr, Vincent Cottin, Sonye K. Danoff, Ferran Morell, Kevin R. Flaherty, Athol Wells, Fernando J. Martinez, Arata Azuma, Thomas J. Bice, Demosthenes Bouros, Kevin K. Brown, Harold R. Collard, Abhijit Duggal, Liam Galvin, Yoshikazu Inoue, R. Gisli Jenkins, Takeshi Johkoh, Ella A. Kazerooni, Masanori Kitaichi, Shandra L. Knight, George Mansour, Andrew G. Nicholson, Sudhakar N. J. Pipavath, Ivette Buendía-Roldán, Moisés Selman, William D. Travis, Simon Walsh, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society



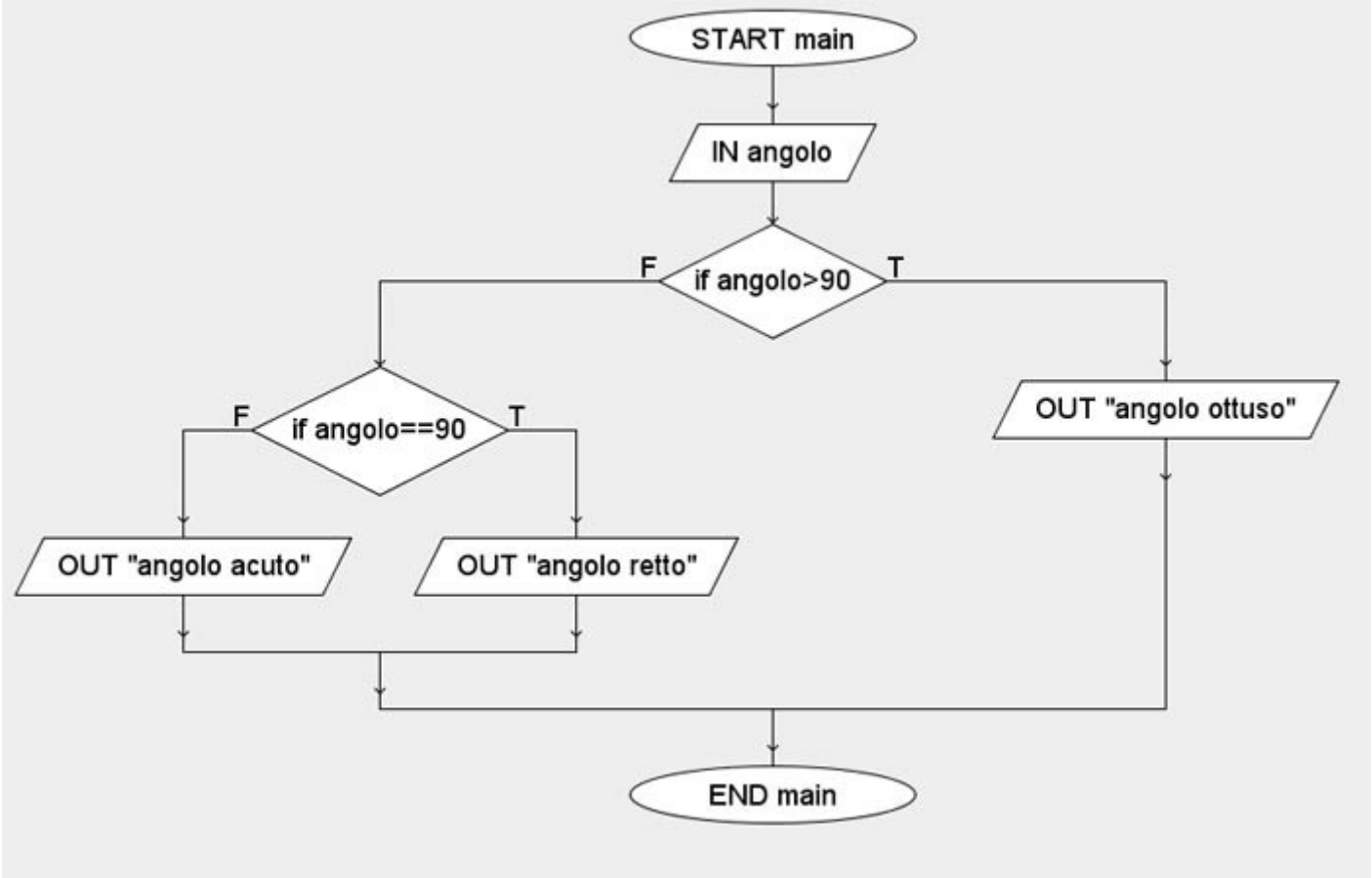
Fisica Classica





Fisica Quantistica

Algoritmo è un procedimento che risolve un determinato problema attraverso un numero finito di passi: elementari, chiari e non ambigui



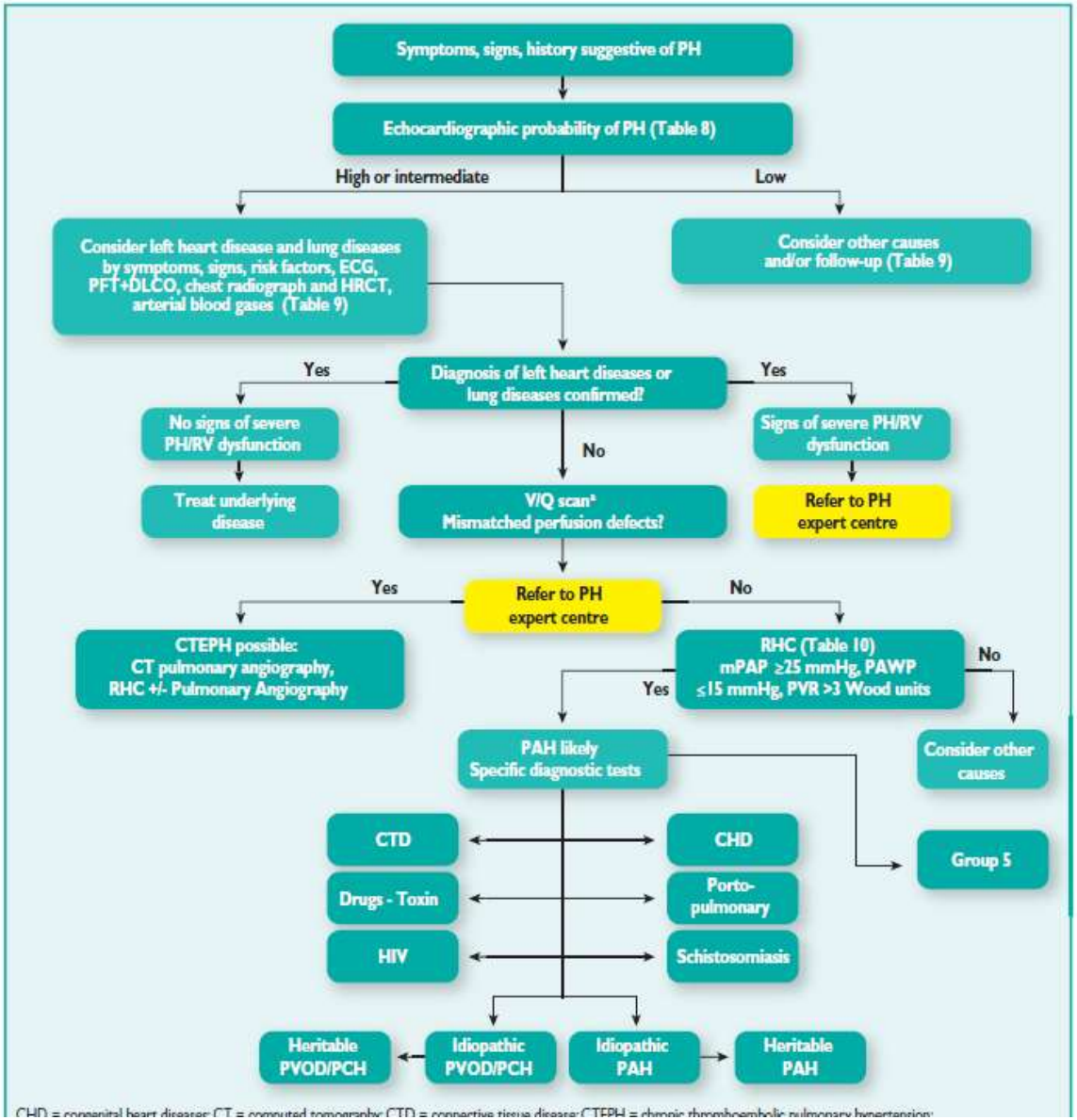
European Heart Journal Advance Access published August 29, 2015

European Heart Journal
doi:10.1093/eurheartj/ehv317

ESC/ERS GUIDELINES

2015 ESC/ERS Guidelines for the diagnosis and treatment of pulmonary hypertension

The Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS)



American Thoracic Society

1999 **Idiopathic Pulmonary Fibrosis: Diagnosis and Treatment**
International Consensus Statement

THIS JOINT STATEMENT OF THE AMERICAN THORACIC SOCIETY (ATS), AND THE EUROPEAN RESPIRATORY SOCIETY (ERS) WAS ADOPTED BY THE ATS BOARD OF DIRECTORS, JULY 1999 AND BY THE ERS EXECUTIVE COMMITTEE, OCTOBER 1999

IPF is defined as a specific form of chronic fibrosing interstitial pneumonia limited to the lung and associated with the histologic appearance of usual interstitial pneumonia (**UIP**) on surgical (thoracoscopic or open) lung biopsy.

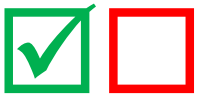
Diagnosi

1999 *In the absence of a surgical lung biopsy, the diagnosis of IPF remains uncertain. However, in the immunocompetent adult…*

all

Major Criteria

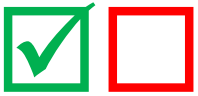
- Exclusion of other known causes of ILD
- Evidence of restriction (reduced VC often with an increased FEV1/FVC ratio) and impaired gas exchange DLCO
- Bibasilar reticular abnormalities with minimal ground glass opacities on HRCT scans
- Transbronchial lung biopsy or bronchoalveolar lavage (BAL) showing no features to support an alternative diagnosis



At least 3

Minor Criteria

- Age >50yr
- Insidious onset of otherwise unexplained dyspnea on exertion
- Duration of illness >3 mo
- Bibasilar, inspiratory crackles (dry or "Velcro" type in quality)

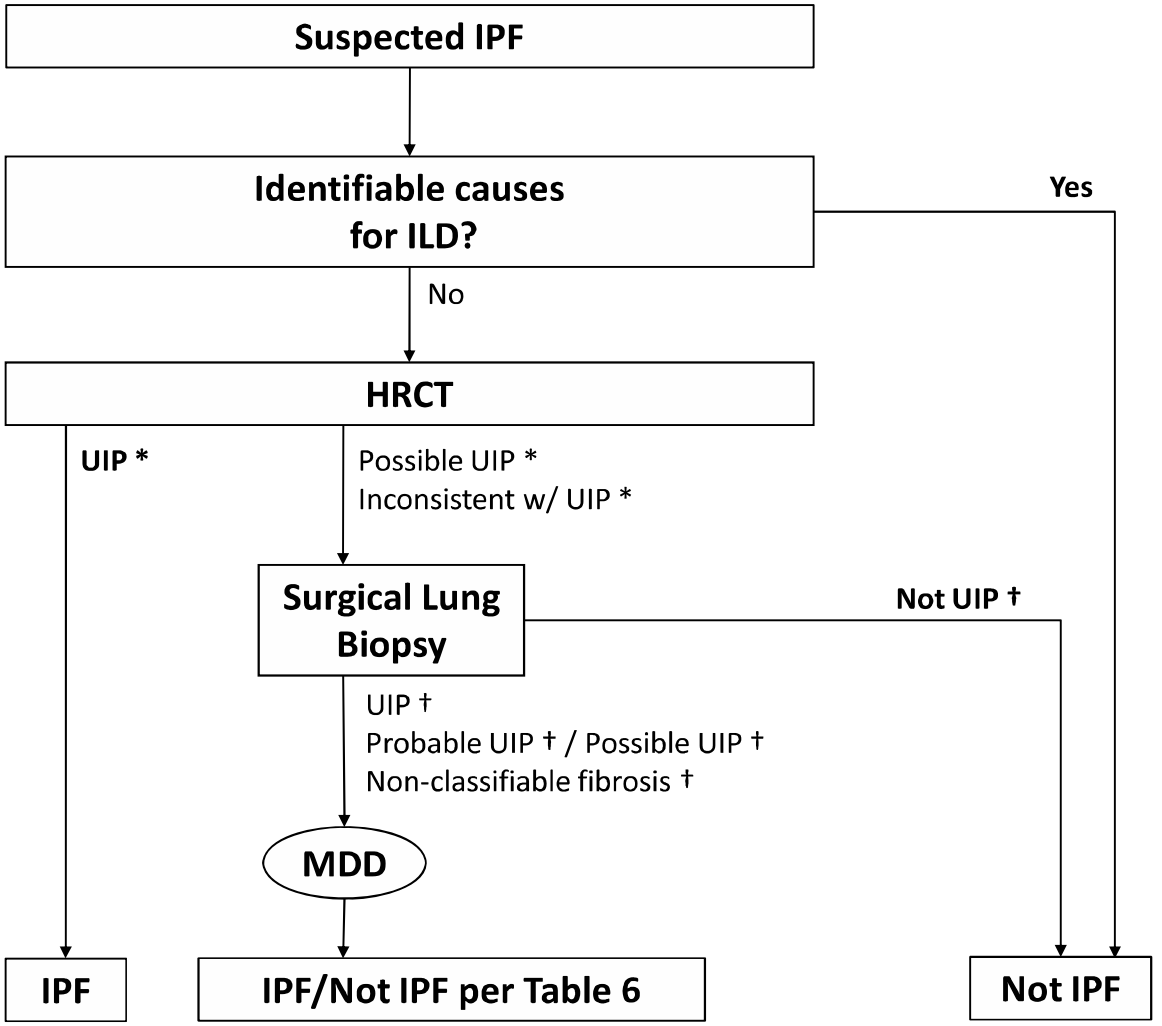


American Thoracic Society Documents

An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management

Ganesh Raghu, Harold R. Collard, Jim J. Egan, Fernando J. Martinez, Juergen Behr, Kevin K. Brown, Thomas V. Colby, Jean-François Cordier, Kevin R. Flaherty, Joseph A. Lasky, David A. Lynch, Jay H. Ryu, Jeffrey J. Swigris, Athol U. Wells, Julio Ancochea, Demosthenes Bouros, Carlos Carvalho, Ulrich Costabel, Masahito Ebina, David M. Hansell, Takeshi Johkoh, Dong Soon Kim, Talmadge E. King, Jr., Yasuhiro Kondoh, Jeffrey Myers, Nestor L. Müller, Andrew G. Nicholson, Luca Richeldi, Moisés Selman, Rosalind F. Dudden, Barbara S. Griss, Shandra L. Protzko, and Holger J. Schünemann, on behalf of the ATS/ERS/JRS/ALAT Committee on Idiopathic Pulmonary Fibrosis

2011



The diagnosis of IPF is multidisciplinary

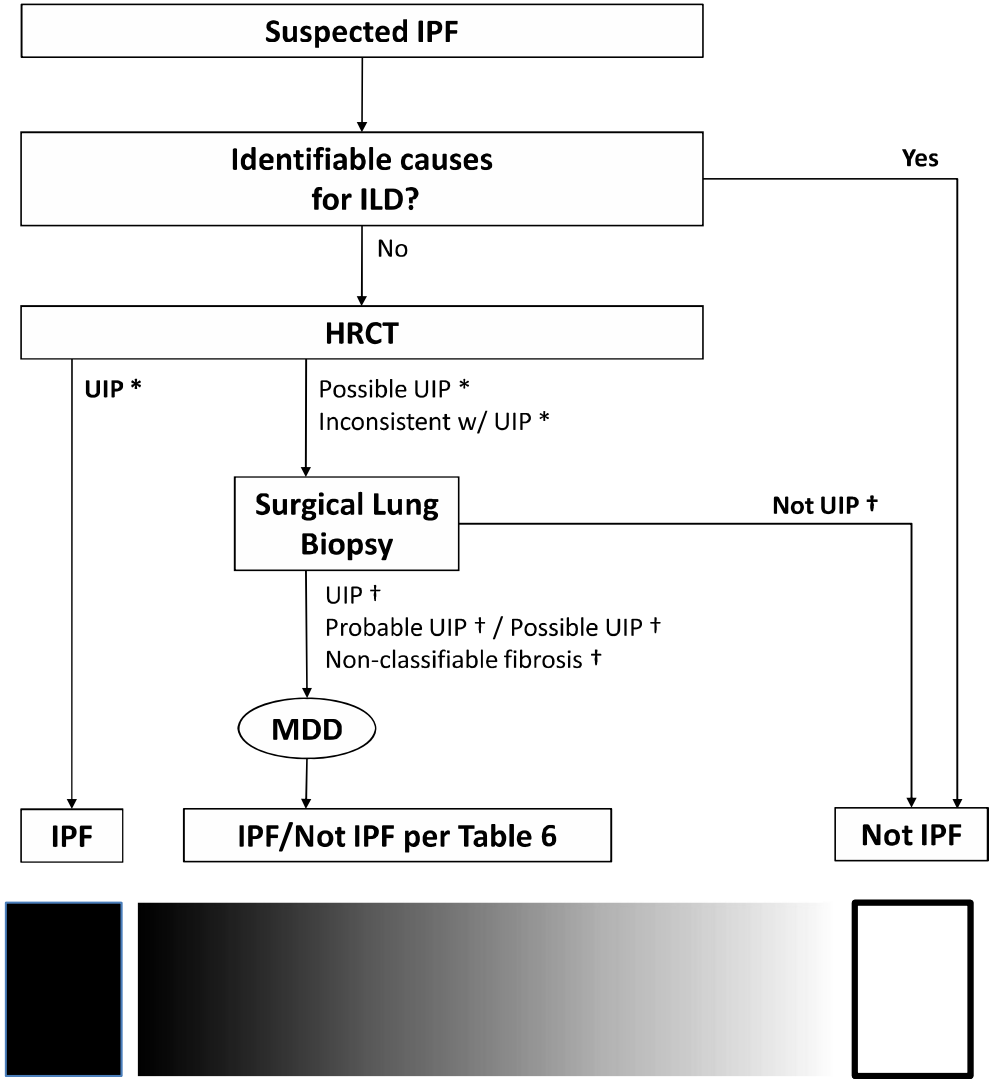
2011

TABLE 4. HIGH-RESOLUTION COMPUTED TOMOGRAPHY CRITERIA FOR UIP PATTERN

UIP Pattern (All Four Features)	Possible UIP Pattern (All Three Features)	Inconsistent with UIP Pattern (Any of the Seven Features)
• Subpleural, basal predominance • Reticular abnormality • Honeycombing with or without traction bronchiectasis • Absence of features listed as inconsistent with UIP pattern (*see* third column)	• Subpleural, basal predominance • Reticular abnormality • Absence of features listed as inconsistent with UIP pattern (*see* third column)	• Upper or mid-lung predominance • Peribronchovascular predominance • Extensive ground glass abnormality (extent > reticular abnormality) • Profuse micronodules (bilateral, predominantly upper lobes) • Discrete cysts (multiple, bilateral, away from areas of honeycombing) • Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes) • Consolidation in bronchopulmonary segment(s)/lobe(s)

TABLE 5. HISTOPATHOLOGICAL CRITERIA FOR UIP PATTERN

UIP Pattern (All Four Criteria)	Probable UIP Pattern	Possible UIP Pattern (All Three Criteria)	Not UIP Pattern (Any of the Six Criteria)
• Evidence of marked fibrosis/ architectural distortion, ± honeycombing in a predominantly subpleural/ paraseptal distribution • Presence of patchy involvement of lung parenchyma by fibrosis • Presence of fibroblast foci • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (see fourth column)	• Evidence of marked fibrosis / architectural distortion, ± honeycombing • Absence of either patchy involvement or fibroblastic foci, but not both • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (*see* fourth column) OR • Honeycomb changes only‡	• Patchy or diffuse involvement of lung parenchyma by fibrosis, with or without interstitial inflammation • Absence of other criteria for UIP (*see* UIP Pattern column) • Absence of features against a diagnosis of UIP suggesting an alternate diagnosis (*see* fourth column)	• Hyaline membranes* • Organizing pneumonia*† • Granulomas† • Marked interstitial inflammatory cell infiltrate away from honeycombing • Predominant airway centered changes • Other features suggestive of an alternate diagnosis


Suspected IPF
Identifiable causes for ILD?
Yes
No
HRCT
UIP *
Possible UIP *
Inconsistent w/ UIP *
Surgical Lung Biopsy
Not UIP †
UIP †
Probable UIP † / Possible UIP †
Non-classifiable fibrosis †
MDD
IPF
IPF/Not IPF per Table 6
Not IPF

Logica Fuzzy

IPF suspected*		Histopathology pattern			
		UIP	Probable UIP	Indeterminate for UIP	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF dx
	Probable UIP	IPF	IPF	IPF (Likely)**	Non-IPF dx
	Indeterminate	IPF	IPF (Likely)**	Indeterminate***	Non-IPF dx
	Alternative diagnosis	IPF (Likely)** /non-IPF dx	Non-IPF dx	Non-IPF dx	Non-IPF dx

Figure 8. Idiopathic pulmonary fibrosis diagnosis based upon HRCT and biopsy patterns.

*"Clinically suspected of having IPF" = unexplained symptomatic or asymptomatic patterns of bilateral pulmonary fibrosis on a chest radiograph or chest computed tomography, bibasilar inspiratory crackles, and age greater than 60 years. (Middle-aged adults [>40 yr and <60 yr], especially patients with risks for familial pulmonary fibrosis, can rarely present with the otherwise same clinical scenario as the typical patient older than 60 years.)

**IPF is the likely diagnosis when any of the following features are present:

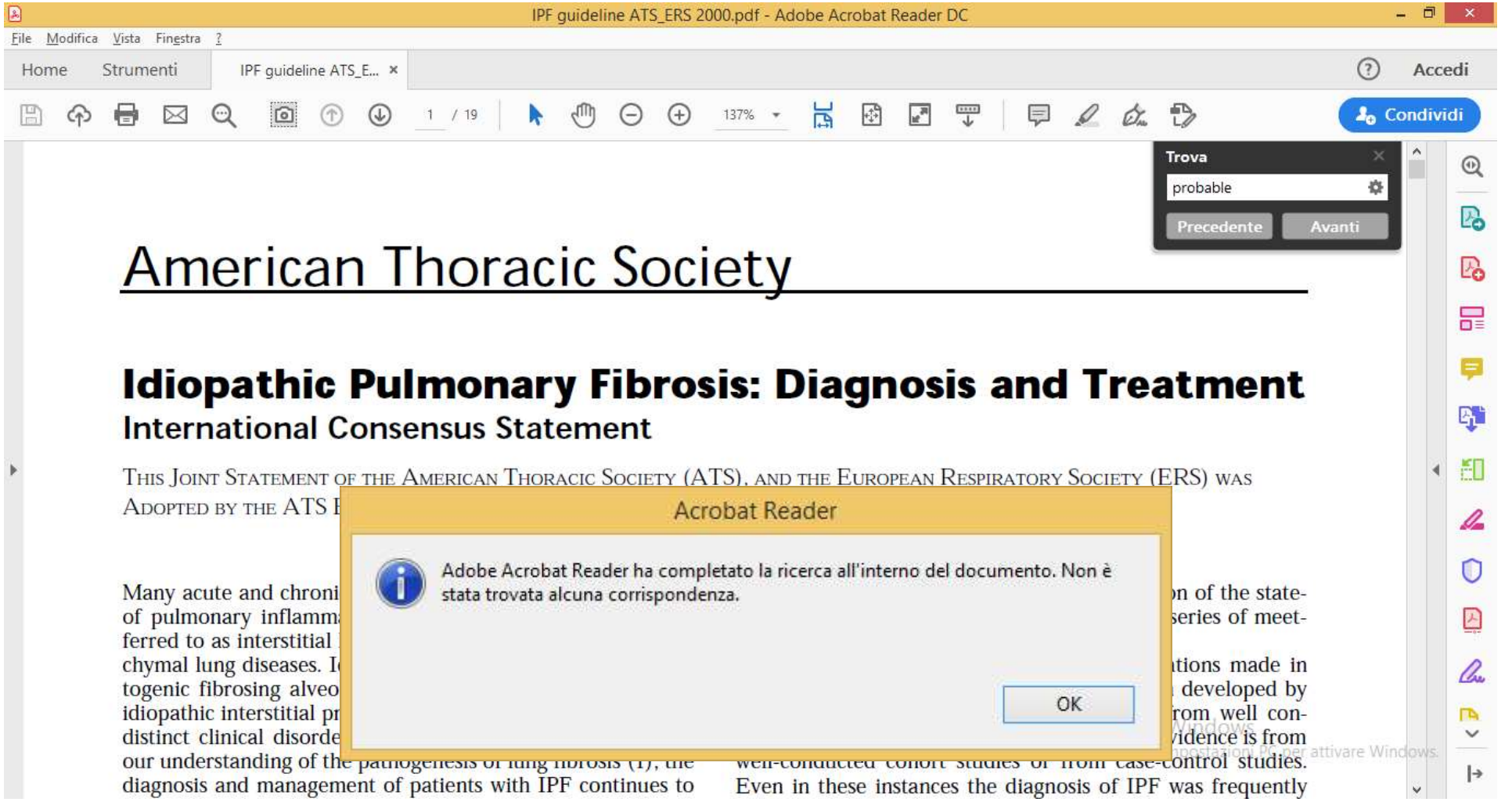
- Moderate-to-severe traction bronchiectasis/bronchiolectasis (defined as mild traction bronchiectasis/bronchiolectasis in four or more lobes including the lingual as a lobe, or moderate to severe traction bronchiectasis in two or more lobes) in a man over age 50 years or in a woman over age 60 years
- Extensive (>30%) reticulation on HRCT and an age >70 years
- Increased neutrophils and/or absence of lymphocytosis in BAL fluid
- Multidisciplinary discussion reaches a confident diagnosis of IPF.

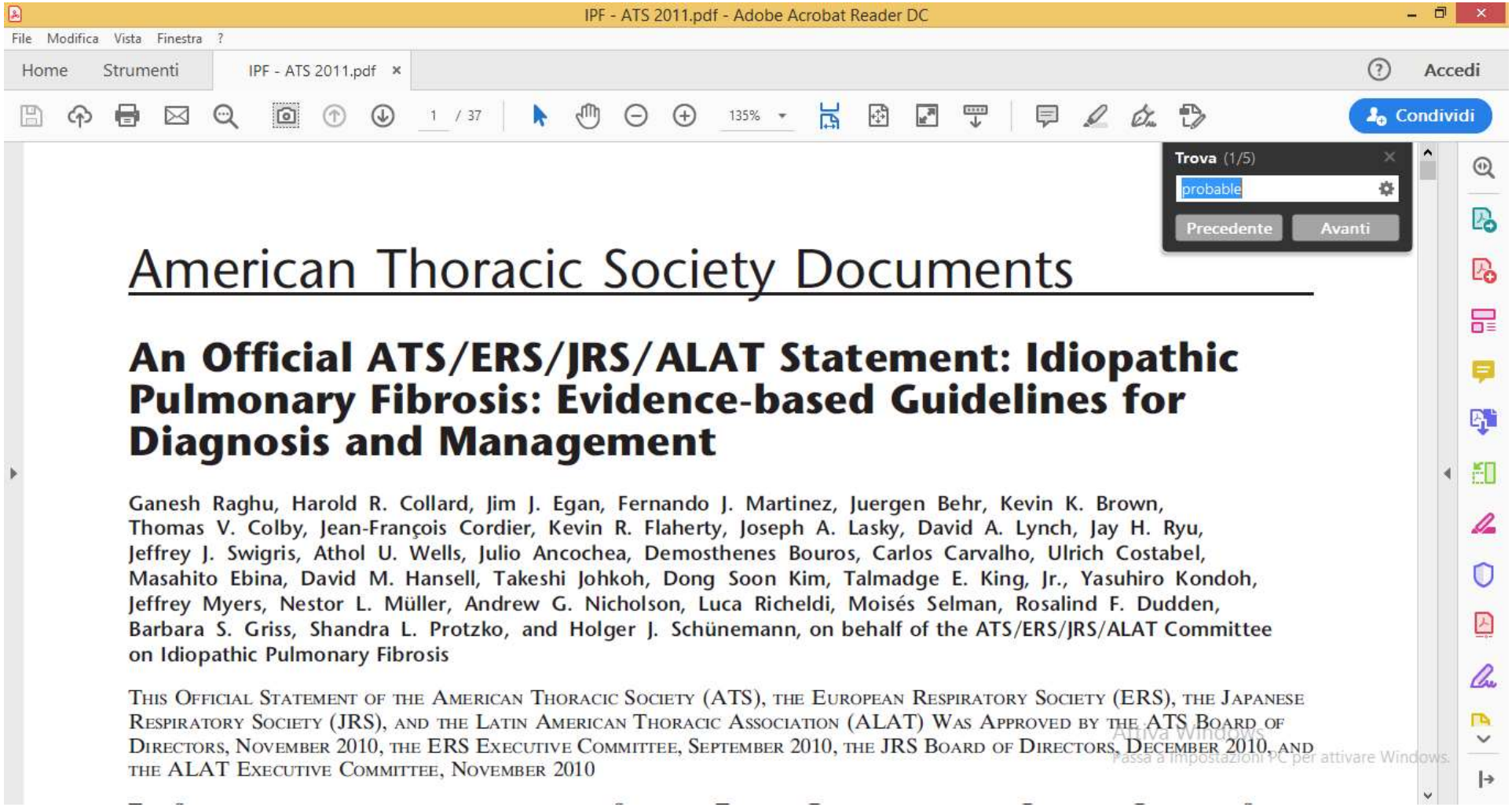
***Indeterminate

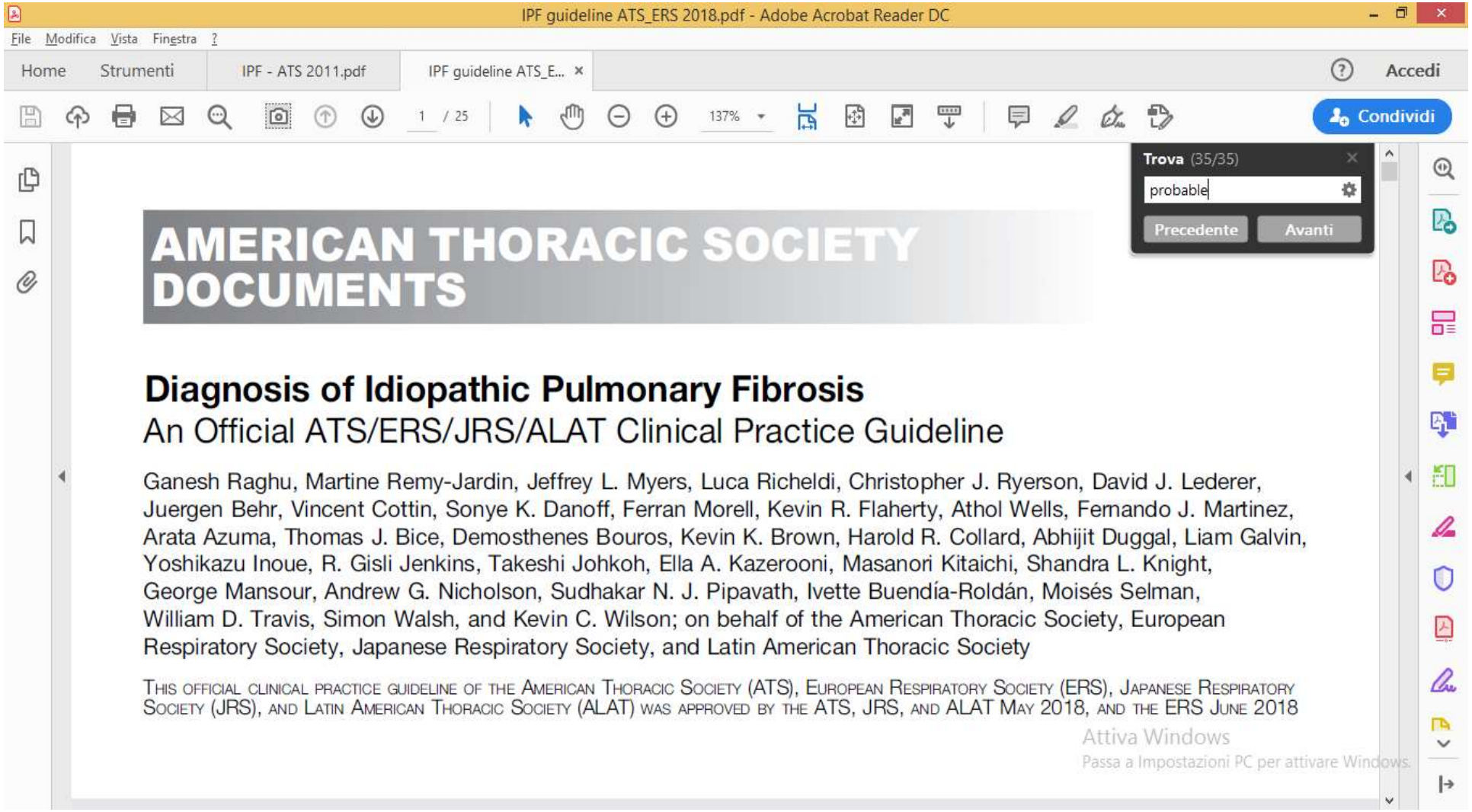
- Without an adequate bio
- With an adequate biopsy iagnosis after multidisciplinary discussion and/or additional consultation.

dx = diagnosis; HRCT = high-res pathic pulmonary fibrosis; UIP = usual interstitial pneumonia.

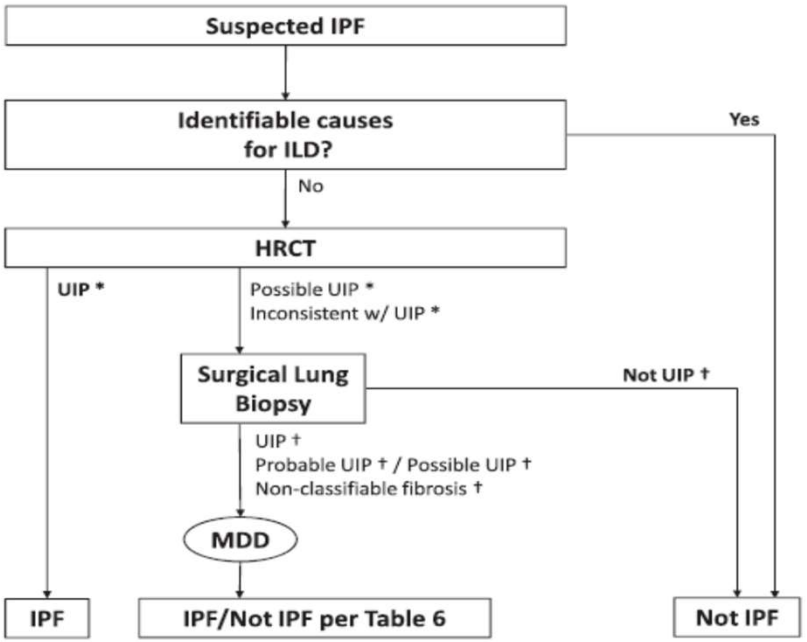
confident



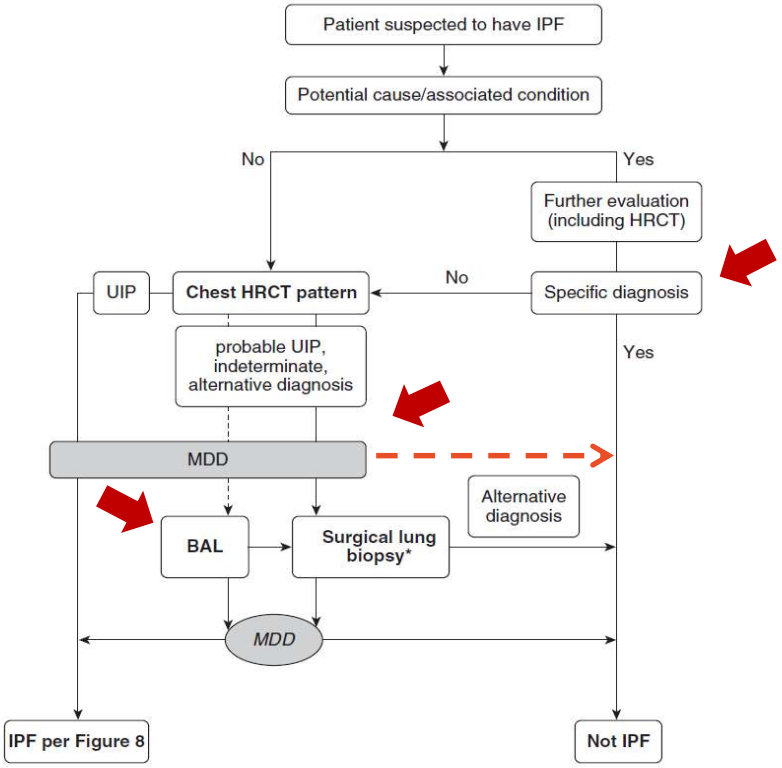


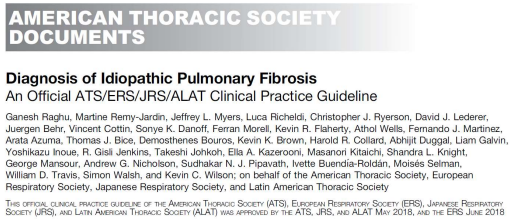


2011


Suspected IPF
Identifiable causes for ILD?
Yes
No
HRCT
UIP *
Possible UIP *
Inconsistent w/ UIP *
Surgical Lung Biopsy
Not UIP †
UIP †
Probable UIP † / Possible UIP †
Non-classifiable fibrosis †
MDD
IPF
IPF/Not IPF per Table 6
Not IPF

2018

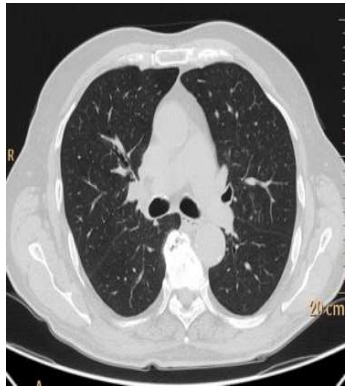

Patient suspected to have IPF
Potential cause/associated condition
No
Yes
Further evaluation (including HRCT)
UIP
Chest HRCT pattern
No
Specific diagnosis
probable UIP, indeterminate, alternative diagnosis
Yes
MDD
Alternative diagnosis
BAL
Surgical lung biopsy*
MDD
IPF per Figure 8
Not IPF



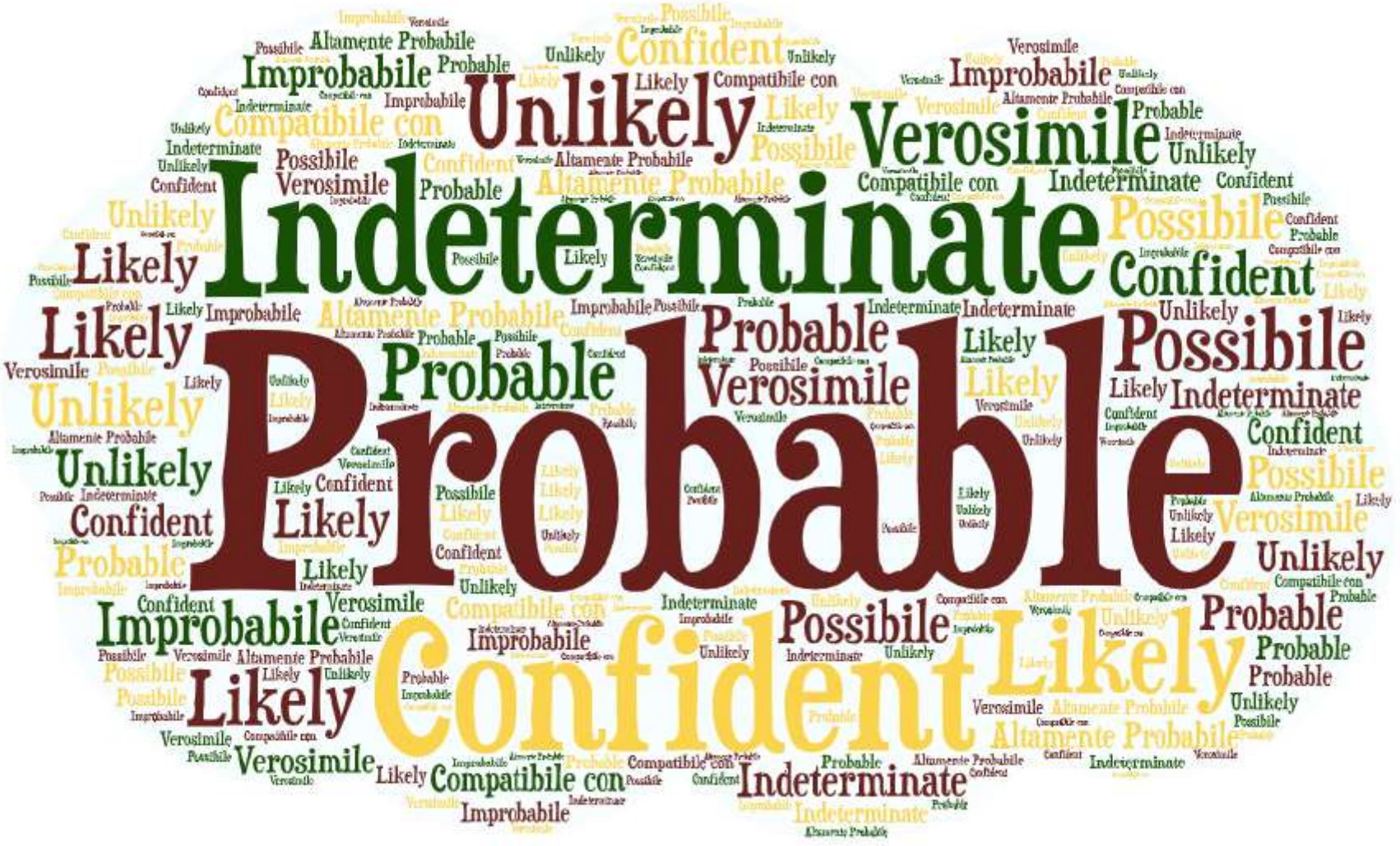
For patients with newly detected ILD of apparently unknown cause who are clinically suspected of having IPF, **we suggest MDD for diagnostic decisionmaking**

"1,000 patients who undergo diagnostic decision-making, SDD and MDD will derive the same diagnosis in **700** patients and different diagnoses in **300** patients. If one accepts MDD as the reference standard, then as many as 300 patients will be potentially subject to incorrect therapy, delayed therapy, or unnecessary additional diagnostic testing"

"For patients with newly detected ILD of apparently unknown cause who are clinically suspected of having IPF, **we suggest MDD for diagnostic decisionmaking**
(*conditional recommendation, very low quality of evidence*)



Esperienza



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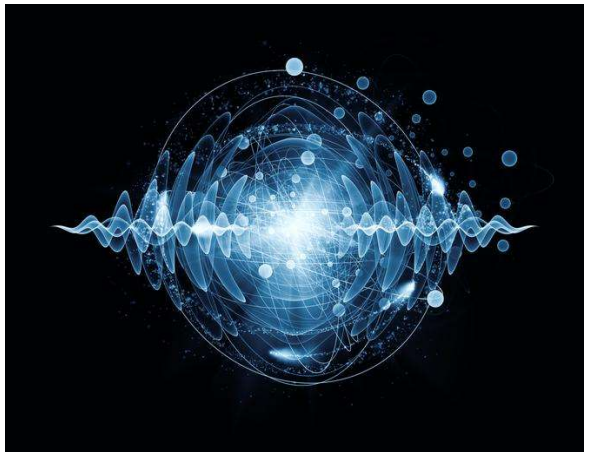
Linee guida ufficiali ATS/ERS/JRS/ALAT per la pratica clinica

1 Febbraio 2020

Hotel Enterprise Milano

SIP Società Italiana di Pneumologia

ITS AIPO



Diagnosi Precoce

Diagnosi Certa