Idiopathic interstitial pneumonias: Classification revision

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The American Thoracic Society (ATS), the European Respiratory Society (ERS) and the Japan Respiratory Society (JRS) are planning a revision of the 2002 ATS/ERS International Multidisciplinary Classification of Idiopathic Interstitial Pneumonias (IIPs)[1]. In two years' time it will be 10 years since its publication and with a view to publishing the revision after 10 years (i.e., in 2012), a steering committee has been established, which met in New Orleans during ATS congress in May 2010 and more recently in Barcelona during the ERS congress (Photo). The committee will meet again during the ATS and the ERS congresses that will be held in the next two years, with an additional meeting in Modena, Italy, in April 2011.

After the introduction of various new classifications over the few past years (Figure 1) it is now an appropriate time to undertake this revision (Figures 2, 3), as the significant advances in every field, including clinical, radiological and pathological, and also the recent genetic data, all need to be incorporated.

The Chair of the committee is the pathologist William D. Travis (Department of Pathology, Sloan Kettering Memorial Cancer Center, New York). Members of the steering committee include clinicians, radiologists, pathologists and molecular physicians (Table 1).

The underlying pathological process is different in each disease entity (Figure 4) and new data on their pathogenesis continue to being accumulated[2-8].

The key issues to be addressed in the classification system include general, clinical, imaging, pathological and molecular questions:

GENERAL QUESTIONS:

- What is the definition of the term IIPs, and is there a better overall term for these entities?
- What are the changes on the longitudinal evaluation of individuals with IIP? How does this affect prognosis?
- Are there any new entities that should be added to the IIP classification or earlier diagnoses that should be removed?
- What approach should be used for cases that are difficult to classify (e.g., overlapping features of usual interstitial pneumonia (UIP) and non-specific pneumonia)?

TABLE 1. International multidisciplinary core committee members.

Pulmonary Medicine	Radiology	Pathology	Molecular
Jurgen Behr, Germany	David Lynch, USA	William Travis, USA	Bruno Crestani, France
Demosthenes Bouros, Greece	David Hansell, UK	Andrew Nicholson, UK	Cory Hogaboam, USA
Kevin Brown, USA	Takeshi Johkoh, Japan	Masanori Kitaichi, Japan	Moises Selman, Mexico
Harold R. Collard, USA	Nicola Sverzellati, Italy	Thomas Colby, USA	James Loyd, USA
Ulrich Costabel, Germany		Jeffrey Myers, USA	
Vincent Cotin, France			
Marjolein Drent, Netherland			
Jim Egan, Ireland			
Kevin Flaherty, USA			
Yoshikazu Inoue, Japan			
Fernando J. Martinez, USA			
Dong Soon Kim, R. Korea			
Talmadge E. King, Jr, USA			
Ganesh Raghu, USA			
Luca Richeldi, Italy			
Carlos Robalo Cordeiro, Portugal			
Jay Ryu, USA			
Dominique Valeyre, France			
Athol Wells, UK			

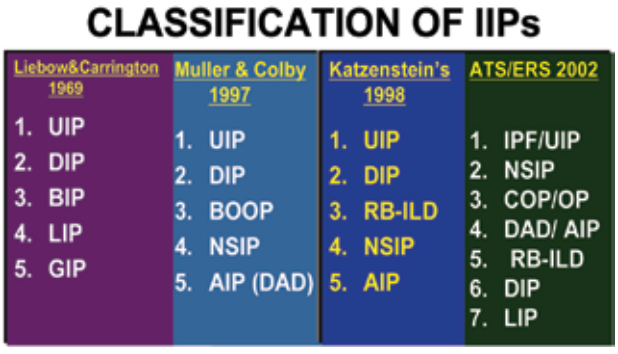


FIGURE 1. The various classifications of idiopathic Interstitial Pneumonias.

- How should imaging/pathological features be reported when the patterns are indeterminate, or mixed?
- What are the less typical findings of UIP on imaging?

CLINICAL QUESTIONS

- Should bronchoalveolar lavage (BAL) findings be used in the diagnosis of IIPs?
- What is the appropriate biopsy for diagnosis of IIPs?
- Does spontaneous clinical improvement or resolution occur in any of the IIPs?

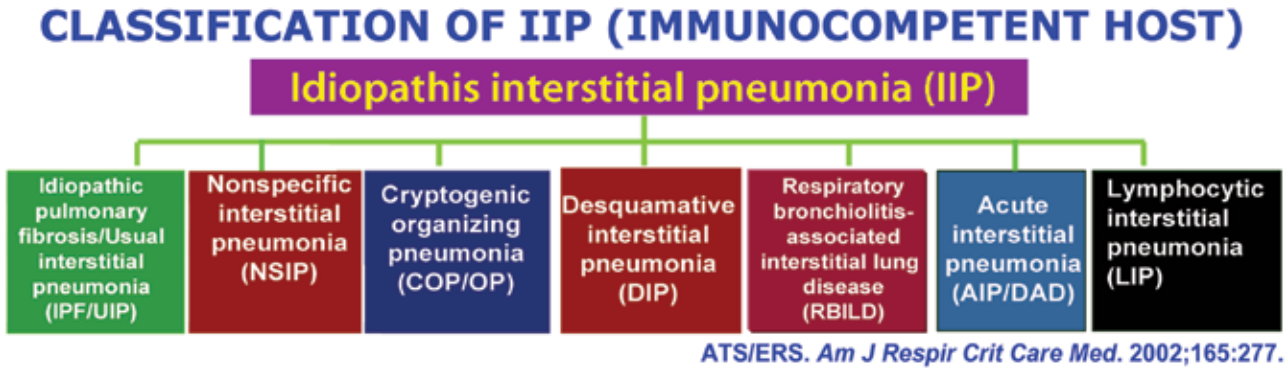


FIGURE 2. Current classifications of idiopathic Interstitial Pneumonias.

UNDERLYING PATHOLOGY OF IDIOPATHIC INTERSTITIAL PNEUMONIAS

CLINICAL CLASSIFICATION	HISTOLOGIC CLASSIFICATION
1. IDIOPATHIC PULMONARY FIBROSIS (IPF)	1. USUAL INTERSTITIAL PNEUMONIA (UIP)
2. NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP)	2. NONSPECIFIC INTERSTITIAL PNEUMONIA (NSIP)
3. CRYPTOGENIC ORGANISING PNEUMONIA (COP)	3. ORGANISING PNEUMONIA (OP)
4. ACUTE INTERSTITIAL PNEUMONIA (AIP)	4. DIFFUSE ALVEOLAR DAMAGE (DAD)
5. DESQUAMATIVE INTERSTITIAL PNEUMONIA (DIP)	5. DESQUAMATIVE INTERSTITIAL PNEUMONIA (DIP)
6. RESPIRATORY BRONCHIOLITIS-INTERSTITIAL LUNG DISEASE (RB-ILD)	6. RESPIRATORY BRONCHIOLITIS-INTERSTITIAL LUNG DISEASE (RB-ILD)
7. LYMPHOCYTIC INTERSTITIAL PNEUMONIA (LIP)	7. LYMPHOCYTIC INTERSTITIAL PNEUMONIA (LIP)

FIGURE 3.

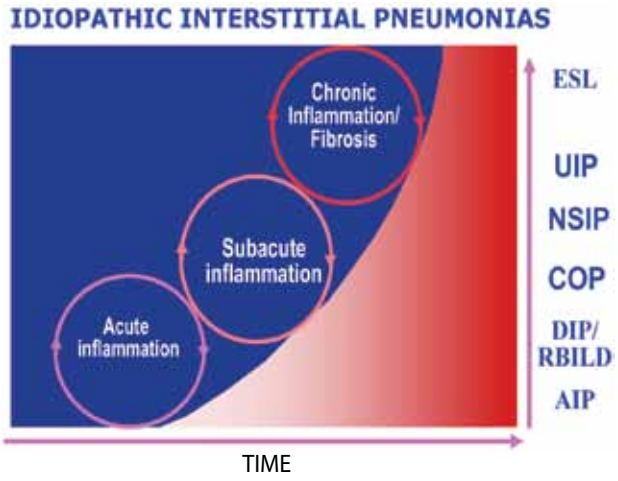
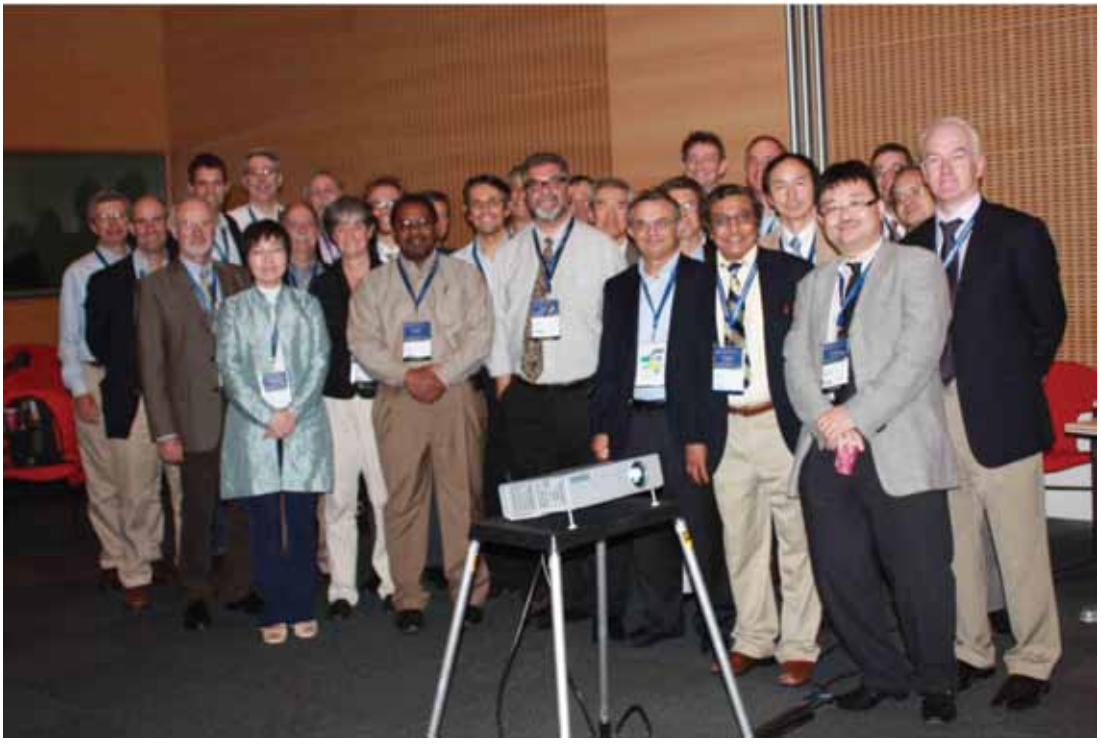


FIGURE 4. Underlying pathology of idiopathic Interstitial Pneumonias.

- How frequent are relapses in patients with chronic obstructive pulmonary disease (COP)? What is the impact of recurrences of COP on the long-term outcome?
- How does the timing of treatment alter the clinical course of patients with COP and the frequency of recurrences?
- Is there any role for biomarkers in the setting of IIPs?



Members of the international multidisciplinary steering committee for the revision of the classification of idiopathic interstitial pneumonias (Barcelona Sept. 2010).

PATHOLOGICAL QUESTIONS

- What are the distinguishing pathological criteria for IIP and are there any changes in the pathological criteria from those used in the 2002 IIP classification?
- What is the pathological definition of honeycombing?
- What terminology should be used to describe the pathological features of IIP?
- How should pathological diagnoses be reported?
  - Should grading of severity be included?
  - Should a fibroblastic scoring system be used in UIP?
  - How should diagnostic levels of certainty be addressed?
- How should problem cases (atypical or mixed patterns) be addressed?
  - What the significance of a granuloma in the setting of IIP pathology?
  - What is the interobserver reproducibility of histological diagnosis for IIP?
  - What is the impact of treatment prior to biopsy on the histological findings?
  - What diagnostic information can be obtained from bronchoscopic/core biopsies for diagnosis of IIP's?

IMAGING QUESTIONS

- What imaging is required to make an accurate diagnosis?
- What is the optimal computed tomography (CT) technique for imaging IIP?
- What terminology should be used to describe the imaging features of IIP?
- What is the radiological definition of honeycombing?
- What are the essential components of an imaging report on a patient with IIP?
- How should the level of diagnostic certainty be addressed?
- What is the role of high resolution CT (HRCT) assessment in the follow-up of patients with IIP and are there settings where this influences classification?
- What is the role of HRCT in selection of the biopsy site?
- In what ways have the imaging criteria changed since the 2002 IIP classification?
- What is the interobserver variation in the imaging diagnosis of IIPs?
- What is the role of PET/CT in diagnosis of IIPs?
- Is there a CT severity index that can be used for staging of IIPs?

MOLECULAR QUESTIONS

- In what specific settings can genetic studies help in the diagnosis of IIP?
- What genetic syndromes are associated with pulmonary fibrosis and what are the radiological/pathological patterns associated with these syndromes?
- How should tissue be managed to provide material for future genetic studies?
- What molecular insights into pathogenesis may help in the concept of classification of IIP?

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