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Opinion

Update of ERS/ATS 2025 Statement the International Multidisciplinary Classification of the Interstitial Pneumonias: What Changes in Clinical Practice?

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Abbreviations

ILDs: Interstitial Lung Diseases; ATS: American Thoracic Society; ERS: European Respiratory Society; UIP: Interstitial Pneumonia; NSIP: Nonspecific Interstitial Pneumonia; DIP: Desquamative Interstitial Pneumonia; OP: Organizing Pneumonia; CTD: Connective Tissue Diseases; HP: Hypersensitivity Pneumonitis; BIP: Bronchiolocentric Interstitial Pneumonia; AIP: Acute Interstitial Pneumonia; DAD: Diffuse Alveolar Damage; PPFE: Pleuroparenchymal Fibroelastosis

Interstitial lung diseases (ILDs) represent a heterogeneous spectrum of pneumopathies affecting the pulmonary interstitium and/or alveolar spaces, with a substantial impact on patient morbidity and mortality. Historically, the classification proposed by the American Thoracic Society (ATS) and the European Respiratory Society (ERS), initially published in 2002 and revised in 2013, marked a milestone in the diagnostic standardization of idiopathic interstitial pneumonias, allowing for comparability between studies, greater diagnostic accuracy, and therapeutic advances [1].

However, over the past 10 years, the growing understanding of ILDs, the incorporation of new clinical, histopathological, and radiological evidence, and the need to integrate secondary causes have led to the publication of the International Multidisciplinary Update of the Classification of Interstitial Pneumonias - ERS/ATS in August 2025 [2].

This new document represents a significant paradigm shift. While the 2013 classification focused mainly on idiopathic forms, the current classification is expanded, refined, and updated to encompass secondary causes of interstitial pneumonias, recognize new patterns, and adjust terminology in conformity with a more accurate understanding of pathogenesis.

For pulmonologists, rheumatologists, radiologists, pathologists, palliative care specialists, and the entire clinical team involved in the complex care of these patients, understanding these changes is not merely an academic exercise but a practical necessity: diagnostic concepts, terminology, and radiological patterns directly influence clinical decision-making, prognosis, and the timely and appropriate selection of treatments, including the increasingly widespread use of antifibrotic agents in different scenarios of progressive pulmonary fibrosis.

What is new?

The 2025 ERS/ATS update on the classification of interstitial pneumonias introduces four key changes with direct implications for clinical practice.

Expansion beyond idiopathic forms to include secondary causes

Until 2013, the classification of ILDs focused exclusively on patterns of idiopathic forms, such as usual interstitial pneumonia (UIP), nonspecific interstitial pneumonia (NSIP), desquamative interstitial pneumonia (DIP), and organizing pneumonia (OP) [1]. Although the 2013 document acknowledged entities such as pneumonia associated with connective tissue diseases (CTD) or hypersensitivity pneumonitis (HP), they were not formally integrated into the classification.

The 2025 document breaks this barrier and integrates idiopathic and secondary forms into a single classification framework, recognizing that: many cases initially classified as idiopathic may later reveal underlying causes (e.g. environmental exposure, drug-induced pneumotoxicity, or autoimmune disease); conditions such as respiratory bronchiolitis associated with ILD or even IPF may, in certain contexts, have external triggers related to identifiable exposures or genetic predispositions; the integration of secondary causes provides future flexibility to incorporate new etiological factors [2].

This means that diagnostic reasoning should begin with histopathological and radiological patterns, as we do in practice today, but always integrated with clinical, environmental, and laboratory investigations to identify potential secondary causes. This approach favors more individualized therapeutic decisions, as well as clearer prognostic implications, such as avoiding relevant exposure to mold or birds in patients suspected of having chronic HP.

Inclusion of new patterns in the classification of ILDs and updates in nomenclature

The revised classification of interstitial pneumonias incorporates newly defined interstitial pneumonias patterns and refines terminology to enhance semantic precision and integrate clinical, radiological, and pathological features.

Bronchiolocentric Interstitial Pneumonia (BIP) is now recognized as one of the main patterns in the ILD classification [2]. Until the 2013 version, bronchiolocentric fibrosis and peribronchiolar inflammatory infiltrates were described only as secondary or atypical manifestations of other ILDs, particularly in chronic HP and aspiration-related lung disease [1]. With its consolidation as a defining pattern, BIP is now positioned alongside the other fibrotic interstitial pneumonias, establishing both a conceptual and practical parallel.

In radiology, BIP typically presents with centrilobular ground-glass opacities, mosaic attenuation, and air trapping [3]. From an a pathological perspective, it is characterized by peribronchiolar inflammation or fibrosis, possibly associated with non-necrotizing granulomas or bronchiolar metaplasia. The relevance of this change lies in distinguishing morphological pattern from clinical diagnosis: while HP should be understood as a multidisciplinary diagnosis requiring thorough exposure investigation [4], BIP is considered a pattern that can be present in different conditions, such as HP, CTD, aspiration, or drug-induced pneumotoxicity.

Another key modification is the replacement of the term acute interstitial pneumonia (AIP) with idiopathic diffuse alveolar damage (DAD). The previous designation was imprecise, as several interstitial pneumonias can present themselves acutely [2]. The proposed terminology provides a more accurate representation of the underlying histopathological substrate, corresponding to diffuse alveolar injury [5]. It correlates directly with the clinical picture of severe acute respiratory failure, similar to acute respiratory distress syndrome [6].

The term desquamative interstitial pneumonia (DIP) was also changed to alveolar macrophage pneumonia (AMP). The previous name was misleading, as there is no desquamation of pneumocytes, but rather a diffuse accumulation of intra-alveolar macrophages. The new nomenclature better reflects the pathophysiology of the entity and avoids diagnostic confusion, particularly in smokers, in whom it may coexist with respiratory bronchiolitis [7].

These changes directly impact clinical practice, as they affect radiology reports and histopathological reports. Adoption of the new nomenclature is recommended to ensure standardization and clarity in multidisciplinary discussions among pulmonologists, rheumatologists, pathologists, and radiologists.

Classification of ILDs into fibrotic vs. non-fibrotic

Another relevant innovation is the subdivision of interstitial pneumonias into fibrotic and non-fibrotic categories. Although previous classifications already recognized prognostic differences between progressive fibrotic and acute/subacute patterns the new approach refines this distinction.

The updated classification of ILDs distinguishes patterns according to their fibrotic or non-fibrotic nature, in addition to a separate category of alveolar filling disorders. Fibrotic patterns include UIP, fibrotic forms of NSIP and BIP, as well as pleuroparenchymal fibroelastosis (PPFE). Non-fibrotic patterns encompass the cellular form of NSIP, the inflammatory form of BIP, lymphocytic interstitial pneumonia (LIP), and interstitial pneumonia with diffuse alveolar damage (DAD) in its acute phase. Finally, alveolar filling disorders comprise OP, respiratory bronchiolitis-associated ILD (RB-ILD), AMP, both acute and chronic eosinophilic pneumonia, and pulmonary alveolar proteinosis.

The subclassification mentioned above has substantial prognostic implications and directly influences the therapeutic decision-making process. Patients with fibrotic patterns share an increased risk of progression to progressive pulmonary fibrosis (PPF), regardless of etiology. Recent clinical trials have demonstrated the benefit of antifibrotics in non-IPF PPF, consolidating the concept that clinical behavior (fibrotic progression) should guide intervention. This change enables earlier initiation of disease-modifying therapies, potentially improving patient outcomes.

Degree of diagnostic confidence

An additional relevant improvement is the formal incorporation of levels of diagnostic confidence for each pattern, both in radiology and pathology, and their integration into multidisciplinary decision-making. The proposed framework stratifies diagnostic certainty into three categories: confident diagnosis (≥90%), provisional diagnosis (51-89%), and unclassifiable disease (<50% confidence in any main hypothesis).

This change promotes greater transparency and standardization in communication among specialists in a complex context where no pathognomonic test exists for most ILDs. Diagnosis is thus built by integrating clinical, laboratory, radiological, and, in some cases, pathological data. The level of confidence should be explicitly stated in reports to guide management, particularly regarding the need for biopsy or initiation of antifibrotic or immunosuppressive therapy.

Conclusion

The 2025 ERS/ATS update to the classification of interstitial pneumonia represents a paradigm shift, expanding beyond idiopathic forms to encompass secondary causes, incorporating new pathological patterns and revised terminology, introducing a clinically meaningful subdivision into fibrotic and non-fibrotic categories, and formally integrating levels of diagnostic confidence into multidisciplinary practice.

Collectively, these changes aim to strengthen clinico-radiological-pathological correlations, improve diagnostic precision, and provide a framework that more directly informs therapeutic decision-making. By aligning nomenclature with pathogenesis, refining prognostic stratification, and guiding the timely initiation of disease-modifying therapies, this update promotes greater standardization in clinical practice and enhances the potential for individualized patient care across the heterogeneous spectrum of ILDs.

Conflict of Interest

R. Figueiredo reports payment or honoraria for lectures, presentations, manuscript writing, or educational events from Ache, AstraZeneca, Boehringer Ingelheim, GSK, Sanofi, and Sun Pharma. The remaining authors have no potential conflicts of interest to disclose.

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