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Eur Respir J 2016; 47: 1293–1295 | DOI: 10.1183/13993003.02084-2015 | Copyright ©ERS 2016

Interstitial pneumonia with autoimmune features: the new consensus-based definition for this cohort of patients should be broadened

From the authors:

We appreciate the careful reading and concerns expressed by Drs. Collins and Raghu regarding our recently published European Respiratory Society/American Thoracic Society Research Statement on "interstitial pneumonia with autoimmune features" (IPAF). Critical assessment of this document and concept is essential to improving our understanding of this novel classification.

The primary criticism expressed in the letter by Collins and Raghu is that the usual interstitial pneumonia (UIP) pattern of disease is not included within the morphological domain, thereby excluding patients with UIP pattern from the IPAF definition. This is an incorrect conclusion that stems from confusion over the role of domains in the IPAF definition. In the IPAF statement, we explicitly state that patients with UIP can be classified as having IPAF. To paraphrase, having a radiological or histopathological UIP pattern does not exclude IPAF but, unlike non-specific interstitial pneumonia, organising pneumonia or lymphocytic interstitial pneumonia patterns, there is no morphological "credit" associated with the UIP pattern. Thus, to be considered as having IPAF, a patient with a UIP pattern on high-resolution computed tomography or by surgical lung biopsy would need to have at least one feature from the other two domains (clinical and serological) or another pulmonary morphologic feature (*e.g.* unexplained multi-compartment involvement or histopathologic evidence of interstitial lymphoid aggregates with germinal centres and/or diffuse lymphoplasmacytic infiltration). We appreciate the opportunity to clarify this point.

A second criticism expressed was that the clinical features within the IPAF classification scheme are too limiting and that features such as oesophageal dysmotility and myalgias should also be included. This is a valid criticism. We acknowledge in our discussion, "… in the absence of data to inform decision-making, we were left to devise what this panel believes to be a reasonable first draft of criteria that can be readily applied by investigators who wish to study this interesting, and presently poorly defined, group of patients."

Finally, Collins and Raghu suggest that the term "autoimmune ILD" is more appropriate for this condition. As was explained in our document, the Task Force believed it was important to use straightforward

nomenclature to describe an interstitial pneumonia that has certain clinical, serological and/or pulmonary morphological features suggesting the presence of an autoimmune process. We believe the term IPAF achieves this goal.

We are optimistic that the proposed IPAF definition and classification allows for the future study of a uniform cohort of patients with interstitial pneumonia and features of an underlying autoimmune disease. The comments of Collins and Raghu provide an opportunity for us to re-emphasise that prospective studies are needed to validate and refine the proposed criteria, and to determine the natural history and clinical implications of such a diagnosis.

@ERSpublications
We clarify that patients with UIP can indeed be classified as interstitial pneumonia with autoimmune features (IPAF) http://ow.ly/YGKLl

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Received: Jan 04 2016 | Accepted: Jan 20 2016

Disclosures can be found alongside the online version of this article at erj.ersjournals.com

Eur Respir J 2016; 47: 1295–1296 | DOI: 10.1183/13993003.00019-2016 |