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Editorial

Updates in the New Guidelines for Idiopathic Pulmonary Fibrosis: Role of Cryobiopsy

Novedades en la nueva guía de la FPI: papel de la criobiopsia

The first guide on the management of idiopathic pulmonary fibrosis (IPF), published in 2000, has been revised and updated on three occasions, and each time, the interval has been shorter; the guidelines are also being newly incorporated by scientific societies. Both facts highlight the growing interest in a high-impact disease and the important advances in its research.[1–3]

The new guide published in May of this year and promoted by the American Thoracic Society, the European Respiratory Society, the Japanese Respiratory Society and the Latin American Thoracic Association (ATS/ERS/JRS/ALAT) continues to use the GRADE methodology, as in previous versions, in which pulmonologists have a prominent role. This version also includes the participation of patient representatives.[4]

Relevant issues have been addressed, mainly focusing on four points: 1. updating the histological-radiological criteria necessary for disease diagnosis, 2. the usefulness of genomic markers, 3. anti-reflux approaches, and 4. the usefulness of transbronchial lung cryobiopsy (TBLC) and its implementation in the diagnostic algorithm, which may have special significance for pulmonologists involved in the management of IPF.

In relation to the histological–radiological criteria, disease diagnosis based on high-resolution thoracic CT findings and on the histological model of four patterns (UIP, probable UIP, indeterminate and alternative diagnosis) remains the same as in the previous guide and in the guide published by the Fleischer Society in the same year.[5] The histological criteria were reviewed and confirmed, and the radiological criteria were slightly modified.

In the radiological pattern section that suggests an alternative diagnosis, “subpleural sparing” has been included (in its presence, clinicians are recommended to consider nonspecific interstitial pneumonia or interstitial pneumonitis related to tobacco), and “extensive lymph node enlargement” has been excluded. In addition, for each radiological pattern, the level of agreement of the radiological UIP pattern (UIPr) with the “histological” UIP pattern (UIPh) is specified (>90% for UIP; 70–89% for probable UIP, 51–69% for indeterminate, and <50% for alternative diagnosis); notably, in the appropriate clinical context, histological confirmation is not required if the radiological pattern is UIP or probable UIP. In the 2018 guidelines, the diagnosis of IPF was only established after excluding a known cause of interstitial lung disease (ILD) and the patient had a high resolution thoracic CT result with a UIP radiological pattern. In the presence of other patterns, a histological sample was needed. This diagnostic approach standardizes the guidelines of the ATS/ERS/JRS/ALAT with the guidelines proposed by the task force sponsored by the Fleischer Society.[5]

Another update in this section is the change in position regarding the finding of a radiological pattern that suggests an alternative diagnosis and a histological model of probable UIP. A non-IPF diagnosis, as termed in the 2018 guidelines, has been reclassified as an indeterminate diagnosis.

Regarding genomic classification, which has a sensitivity of 68% and a specificity of 92% in the diagnosis of a UIP pattern in patients with indeterminate ILD, this edition does not comment with regard to its diagnostic utility.

In the therapeutic section, unlike the previously published guidelines, the main update is the position against both the use of antacids and the referral of patients to anti-reflux surgery.

Regarding the use of TBLC, which is of particular interest to our specialty, an increasing number of review articles and meta-analyses support the usefulness of cryobiopsy, which has a diagnostic performance comparable to that of surgical lung biopsy (SLB) according to a multidisciplinary committee, even when the histological diagnostic rate is somewhat higher in samples obtained invasively (80% versus 95% respectively).[6–9] At the same time, the first works have been published to standardize the performance of the procedure to minimize complications, which are less frequent than complications associated with SLB.[10,11] In this update of the guide, given that more scientific evidence is needed because it is a relatively new technique, TBLC is positioned as an acceptable alternative to SLB for the first time as long as it is performed in centres with experience in the endoscopic technique and in the histological analysis of such samples.

Spanish pulmonologists have played a prominent role in the development of these guidelines and their review. In all editions, except the first, the committee included a Spanish pulmonologist as an expert in the field. In the diagnostic section, since the first study assessing the usefulness of TBLC in the diagnosis of ILD published in 2009 and the first national study in 2010, numerous studies have been published, with some carried out in different centres in our country; some of these were pioneering studies because they were published when the actual usefulness of the technique was still questioned.[12] Unsurprisingly, of the 40 articles that have been

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reviewed in this guide to evaluate the usefulness of TBLC in the diagnosis of IPF, five are from Spain.[13–17] Another aspect to consider is that the proposed diagnostic algorithm is similar to that proposed by the CRIOMPID working group and published in our journal.[18] If our group in 2016 questioned whether our experience with and results for TBLC placed us on the right path, this update supports those who have continued to provide evidence and incorporate it into the routine activities of multidisciplinary teams during this time.[13]

Therefore, this new version aligns the diagnostic criteria with those proposed by the Fleischer Society, provides a valuable strategy for the diagnosis of the disease in which TBLC has acquired an increasingly relevant role, positions the value of patient care, and highlights the increasingly notable role of Spanish pulmonologists in understanding this pathology, serving as a stimulus to continue and promote new research plans in this field.

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Conflicts of interest

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Jose Antonio Cascante Rodrigo*, Pilar Cebollero Rivas
Pneumology Service, University Hospital of Navarre (Hospital Universitario de Navarra), Public University of Navarre (UPNA), Spain

* Corresponding author.
E-mail address: casro@separ.es (J.A. Cascante Rodrigo).