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Inherent weaknesses of the current ICD coding system regarding idiopathic pulmonary fibrosis

To the Editor:

Idiopathic pulmonary fibrosis (IPF) is the most prevalent of the idiopathic interstitial pneumonias (IIPs). It carries an ominous prognosis with a median survival of 3 years. Its epidemiology is poorly described because of its rarity and lack of unanimity in diagnostic and coding practices [1]. However, during the last few years, significant improvement has been achieved in our understanding of the pathogenesis, diagnosis

and management of IPF and IIP in general. This is reflected in the 2011 American Thoracic Society (ATS)/European Respiratory Society (ERS)/Japanese Respiratory Society/ALAT consensus statement on IPF [2] and the 2013 ATS/ERS update of the International Multidisciplinary Classification of the Idiopathic Interstitial Pneumonias [3]. To obtain a robust understanding of the epidemiology of IPF is important, especially as novel therapies are emerging. The International Classification of Disease (ICD) coding system represents a great opportunity to create such registries that will expand our knowledge on IPF epidemiology. However, under its current formation, the 10th edition (ICD-10) coding system has severe deficiencies regarding the accurate classification of IPF.

The purpose of the ICD is to permit the systematic recording, analysis, interpretation and comparison of mortality and morbidity data collected in different countries or areas and at different times. However, it is acknowledged that although it is suitable for many different applications, it does not serve all the needs of its various users. It does not provide sufficient detail for some specialties and, sometimes, information on different attributes of health conditions [4]. IPF is such an example. In particular, ICD-10 classifies IPF under the term “other interstitial pulmonary diseases with fibrosis” (J84.1). This category includes diffuse pulmonary fibrosis, fibrosing alveolitis (cryptogenic), Hamman–Rich syndrome, IPF and usual interstitial pneumonia (UIP) [4]. Given the current classification, some problems are readily evident, as follows.

- 1) In the most recent ATS/ERS update on the classification of IIPs, the term “cryptogenic fibrosing alveolitis” was removed, thus leaving IPF as the sole clinical term for this diagnosis [2]. The reason for this was to avoid ambiguity in the nomenclature and definition of the disease.
- 2) “Hamman–Rich syndrome” is an obsolete term that corresponds to acute interstitial pneumonia (AIP). AIP can complicate the natural course of IPF but it is a distinct clinical entity. It is frequently confused with other clinical entities characterised by rapidly progressive interstitial pneumonia, especially secondary acute interstitial pneumonia, acute exacerbations and accelerated forms of IPF. Furthermore, many authors use the above terms both erroneously and interchangeably [5].
- 3) UIP is a radiological and histological pattern. While UIP is the pathological background of IPF, it is not synonymous with it. Other clinical entities with a different prognosis can also be associated with UIP (e.g. chronic hypersensitivity pneumonitis, collagen vascular disease, pulmonary drug toxicity, asbestosis, familial IPF and Hermansky–Pudlak syndrome).
- 4) Finally, the term “diffuse pulmonary fibrosis” is a generic term that does not correspond to any specific clinical or histological entity. This carries the risk of becoming a “bucket” that will include cases of unspecified pulmonary fibrosis irrelevant to IPF. As interstitial lung diseases (ILDs) cover >200 different disease entities, this can lead to obvious misclassification. In our centre, >50% of cases with a final diagnosis of IPF were initially referred as (unclassifiable) pulmonary fibrosis. Thus, based on our personal experience, ICD-10 can lead to false epidemiological data.

We suggest that the upcoming ICD should adopt the classification system proposed by the updated new IIPs classification of the ERS/ATS [3]. Specifically, we suggest the following.

- 1) Each one of the six major IIPs (IPF, idiopathic nonspecific interstitial pneumonia (NSIP), respiratory bronchiolitis–ILD, desquamative interstitial pneumonia, cryptogenic organising pneumonia and AIP) should receive a specific code.
- 2) A separate code should be designated for unclassifiable pulmonary fibrosis. However, this diagnosis should not be considered an easy solution. We recommend that a diagnosis of unclassifiable pulmonary fibrosis should be restricted to expert centres where the diagnostic approach within the context of multidisciplinary discussion according to current guidelines is feasible [2, 6].
- 3) Regarding rare clinical entities (acute fibrinous and organising pneumonia, and idiopathic pleuroparenchymal fibroelastosis), given their rarity, we do not think that each one should be classified under a separate code but under the term “other rare specified interstitial pulmonary diseases”. We suggest that lymphocytic interstitial pneumonia should be categorised in the context of lymphoproliferative disorders (the justification for this is beyond the scope of this correspondence).
- 4) It is not yet clear whether combined pulmonary fibrosis emphysema represents a distinct clinical phenotype with a different prognosis. So far, studies have reached different conclusions regarding mortality [7, 8]. Therefore, we recommend that it should not be classified as a distinct clinical entity but according to the fibrotic disease with which it is observed (e.g. UIP, NSIP or collagen tissue-related ILD).

Currently, after years of negative clinical trials and disappointment, there is effective medication for IPF patients [9–12]. Besides pharmaceutical management, genetic studies and preventive and regenerative strategies (including stem cell transplantation research [13]) exhibit hopeful results. In order to evaluate the impact of the new therapeutic approaches, it is important to have valid data regarding the incidence,

prevalence and mortality of IPF at both local and international levels. An updated ICD based on the current ATS/ERS classification of the IIPs can be a vital tool for achieving this goal.

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ICD-10 does not adequately classify IPF <http://ow.ly/HHNqD>

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