J Bras Pneumol. 2017;43(6):401-402
http://dx.doi.org/10.1590/S1806-37562017000060003

Idiopathic pulmonary fibrosis in Brazil: challenges for epidemiological characterization and management

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Idiopathic pulmonary fibrosis (IPF) is characterized as a progressive fibrosing disease of variable evolution, restricted to the lungs, and classified within the group of idiopathic interstitial lung diseases.[1,2] Although great advances have been made in recent years, which have allowed a better understanding of the pathophysiology of the disease, as well as the determination of more restricted diagnostic criteria and the approval of two antifibrotic medications (pirfenidone and nintedanib) for its treatment, various related aspects need to be optimized, especially in Brazil.[1,5]

In the current issue of the JBP, Algranti et al. reported an increase in IPF-related mortality in Brazil between 1979 and 2014.[6] Although their results reflect the global trend and are quite interesting, due to the long evaluation period, there are some points that deserve to be highlighted. As discussed by the authors, some factors may explain the results found, such as the increase in the life expectancy of the population and the increase in the availability of HRCT, the increase in the number of patients and the greater specificity of the diagnosis of the disease being determined in recent years. In addition, the more restrictive criteria for the diagnosis of IPF were defined in 2000 and readjusted in 2011.[1,7] In this context, the evaluation of the data in the initial period of the study might have caused that various patients with IPF to be misdiagnosed; in fact, those patients might have had other fibrosing lung diseases that surely have a better prognosis than does IPF, which might have contributed to underestimating mortality in the earliest stages. This is reinforced by the fact that approximately 10% of the deaths occurred in individuals under 50 years of age, the diagnosis of IPF being less likely in this age group. Therefore, the disease mortality has certainly increased in recent years; however, that rate might be overestimated.[6]

As described in a recently published article, robust data on the incidence, prevalence, and mortality of IPF in Brazil are yet to be determined, which, in addition to their epidemiological importance, are economically important so that the costs to provide the drugs that have been approved for the treatment of the disease can be estimated.[8] Therefore, it is essential that future studies should be conducted in order to determine the epidemiological data of IPF in our country in a more reliable way, especially from the moment when the diagnostic criteria were better established. In this context, the creation of a Brazilian national case registry database for IPF is urgently needed.

The number of referral centers for the treatment of interstitial lung diseases is still scarce in Brazil. In those centers, multidisciplinary discussions involving pulmonologists, surgeons, radiologists, and pathologists are essential to optimize the diagnostic approach, since several fibrosing lung diseases take part in the differential diagnosis of IPF, such as hypersensitivity pneumonitis (HP) and connective tissue diseases, among others.[1,9-12]

In a recent study by Lynch et al.,[13] a new tomographic classification for IPF was presented. In accordance with that classification system, probable usual interstitial pneumonia was characterized by the presence of reticular opacities and traction bronchiolectasis, with basal and peripheral predominance, no honeycombing, and no other characteristics suggestive of an alternative diagnosis. According to those authors, this tomographic pattern has a high predictive value for IPF in individuals over 60 years of age, smokers or former smokers, with no suggestion for other potential causes of interstitial fibrosing lung diseases, such as chronic HP or connective tissue disease, dispensing with lung biopsy for diagnostic confirmation.[13] This new classification system, in which lung biopsy is not mandatory for the diagnostic confirmation of IPF, creates a trend towards an increase in the number of patients diagnosed with the disease, resulting in greater risks for patients with other fibrosing lung diseases, such as chronic HP and fibrotic nonspecific interstitial pneumonia, to be misdiagnosed with IPF. This aspect is extremely relevant in Brazil, considering the high prevalence of chronic HP, even with a tomographic pattern of probable usual interstitial pneumonia, and the indeterminate frequency of IPF in our country; therefore, this new tomographic criterion should be carefully used in our population with fibrosing lung diseases.[9,14]

Recently, two antifibrotic drugs, pirfenidone and nintedanib, have been approved by the Brazilian National Health Oversight Agency for the treatment of IPF in Brazil, since they reduce the functional decline associated with the disease.[2-5] However, there is a need to provide the access to and greater availability of these medications in our country, based on government funding. Another aspect that is a cause for concern in Brazil is the limited number of referral centers for (unilateral or bilateral) lung transplantation, a therapeutic alternative for patients with IPF that will determine an increase in survival, as well as functional improvement and better quality of life in these patients.[1,2,15,16] Other critical factors that influence this issue is the late referral of patients to referral centers for lung transplantation and the shortage of lung donors,

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which has been below the number of candidates on a waiting list.[15]

In summary, despite the evolution in knowledge of the pathophysiology of IPF and its diagnostic and therapeutic approach, there are several aspects that need to be improved notably in Brazil, including: 1) to broaden the information regarding the disease; 2) to create a Brazilian national case registry database in order to try to obtain more robust data on the incidence, prevalence, and mortality of IPF; this database is about to become available soon, and professionals should be aware of the importance of entering their data into the database system; 3) to widen the availability of and the experience with the use of antifibrotic drugs; and 4) to increase the number of referral centers for the multidisciplinary approach to fibrosing lung diseases and lung transplantation. With these improvements, it will be possible to characterize Brazilian national epidemiological data related to the disease more accurately, including the perspective of determining the impact of antifibrotic drugs on mortality.

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