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Examining the incidence of interstitial lung disease subtypes in South America

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Interstitial lung disease (ILD) comprises a heterogeneous group of over 250 disorders that can be broadly categorized as being secondary to connective tissue disease (CTD-ILD), granulomatous parenchymal lung disease (hypersensitivity pneumonitis [HP] and sarcoidosis), occupational pneumoconiosis, drug-induced lung disease, familial pulmonary fibrosis, or idiopathic interstitial pneumonias. The latter group includes idiopathic pulmonary fibrosis (IPF), which is the prototypic type of progressive fibrosis which can lead to respiratory failure and death within 4-5 years. In a small subset of cases no unifying diagnosis can be confirmed, and these are often referred to as unclassifiable ILD. While many of the previous epidemiologic studies have primarily focused on IPF, regional variability in the prevalence and incidence of ILD is historically less well understood. This issue of the *Jornal Brasileiro de Pneumologia* introduces the first English language literature[1] on incidence of ILD in South America, an important contribution to our global understanding of these diseases.

Much of our current understanding of the variability of global epidemiology of ILD was recently summarized in a review that was able to comment on incidence and prevalence of the various subclassifications of ILD from North America, Europe, Asia, the Middle East, and Australia, but not South America. In North America and Europe, IPF and sarcoidosis were the most prevalent disorders, whereas in Asia a higher relative frequency of HP was observed (10.7-47.3% in India, 12.3% in Pakistan). The greatest variability was with the diagnosis of CTD-ILD, which ranged from 7.5% in Belgium to approximately one third of cases in Canada and Saudi Arabia.[2]

Until now, there has been a notable gap in the literature regarding the incidence of ILD in South America and it has been postulated that previous lack of access to CT scanning and specialized pathologist/radiologist assessment has been a contributing factor.[3] However, thanks to registry data available across six national reference centers in Brazil this is no longer the case. Using retrospective review of cases of incident ILD from this registry over six years, the authors are able to describe the relative frequency of the different ILD subtypes in Brazil for the first time. Whereas other studies have included small or single-center populations, their study[1] is strengthened by the large multicenter sample size of over 1,000 patients, assessment of atypical cases by expert multidisciplinary discussion, and a high proportion of cases with available histopathologic data. The population studied showed slight female predominance with a majority of fibrotic ILD (73.7%). The most common ILD diagnosed was CTD-ILD (26.8%) followed by HP (23.2%) and IPF (14.1%). These findings highlight important differences in the ILD population in South America; in particular, the increased incidence of CTD-ILD and HP is more similar to recent studies from India and Saudi Arabia as compared to Europe and North America.[4,5]

Whether differences in reported ILD frequencies represent true ethnic or geographic variability has been difficult to conclude. Studies using registry data are always affected by selection and referral bias, and there have been significant differences in disease classification (reflecting inconsistent or changing diagnostic criteria) and methodologies between studies. One example in this study[1] is that the authors have elected to include ILD with autoimmune features (IPAF) with CTD-ILD, which comprised 14.7% of total cases and contributed to the relatively large overall prevalence of CTD-ILD observed. Another study to have included IPAF in this category was from Saudi Arabia, who reported a similarly increased incidence of 34.8%.[5] As only a small percentage of patients with IPAF progress to a diagnosis of CTD-ILD and management is not standardized,[6] its inclusion with confirmed CTD-ILD is debatable. However, the authors justified it to emphasize that close collaboration with Rheumatology should be encouraged as their input may improve the specificity of diagnosis in this significant cohort of patients.

Similar to CTD-ILD, the incidence of HP secondary to mold and bird/feather exposure was increased in Brazil, which was attributed to housing conditions with damp indoor spaces and an increased number of captive birds being held in close proximity to humans in the region. In other regions with higher frequency of HP, such as India, it is hypothesized that mold from air coolers may also be implicated.[4] By identifying regions with increased HP and their most prevalent culprit antigens, we grow closer to being able to develop regionally specific questionnaires that can be validated and reliably used to identify relevant exposures, something that has previously been called for in the literature.[7]

In summary, establishing the incidence of ILD in Brazil is an important contribution to our global understanding of this subset of diseases and can be used both locally and internationally to inform and influence clinical practice and public health policy. Future efforts to define regional differences in ILD subtypes would benefit from standardization of diagnostic criteria and study methodology to reduce heterogeneity and better elucidate potential ethnic, geographic, and environmental risk factors for ILD.

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