



## How many patients with idiopathic pulmonary fibrosis are there in Brazil?

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### TO THE EDITOR:

Idiopathic pulmonary fibrosis (IPF) is a form of chronic fibrosing interstitial pneumonia of unknown etiology that primarily affects the elderly and for which usual interstitial pneumonia is the substrate.<sup>(1)</sup> Patients with IPF usually experience progressive loss of pulmonary function and severely impaired quality of life, evolving to death.

The treatment of IPF has always been a challenge. However, in late 2014, the US Food and Drug Administration approved two new drugs for the treatment of patients with this disease. Now it is clear that both drugs reduce the rate of decline in pulmonary function in individuals affected by the disease. In addition, clinical trials are underway to investigate new drugs with different mechanisms of action.

We are seeing the beginning of a new era in the care of IPF patients, which is promising but also implies additional needs and concerns. In particular, health-related governmental decisions should be based on robust epidemiological data, and, unfortunately, there are few such data on IPF in Brazil.

One important and still unresolved issue concerns the true incidence and prevalence of IPF and, consequently, the total number of affected patients in Brazil. When we analyzed the international literature, we found that characterizing the epidemiology of IPF is not a problem solely in Brazil. Some of the difficulties stem from the fact that the current definition of the disease came into use from 2000 onward. In addition, results vary depending on the criteria used by different authors to define a case of IPF. Nevertheless, there is a consensus that the disease affects more men than women, that it is more common after the fifth decade of life, and that its incidence has been increasing over the years, as has the associated mortality.<sup>(1)</sup> Whether the increased incidence and mortality rates are due to greater recognition of the disease, increased survival of the population, or environmental factors is an open question.

A recent systematic review suggested, in a conservative estimate, that the incidence of IPF was approximately 3-9 cases/100,000 population in North America and Europe.<sup>(2)</sup> Incidence rates seem to be lower in South America and Asia. Another, slightly earlier, review indicated that the prevalence of IPF in the United States and European countries was 14.0-27.9 and 1.25-23.4 cases/100,000 population, respectively.<sup>(3)</sup> It is reasonable to assume that various age profiles, as well as ethnic and genetic differences among the populations, may substantially contribute to the different findings.

As previously mentioned, information on the subject of IPF is scarce in Brazil. One study analyzed IPF incidence and mortality data available on the *Departamento de Informática do Sistema Único de Saúde (DATASUS, Information Technology Department of the Brazilian Unified Health Care System)* website for the 1996-2010 period.<sup>(4)</sup> There were progressive increases in both parameters during that period.<sup>(4)</sup> In 2010, the recorded incidence of IPF in Brazil was 4.48 cases/1,000,000 population, whereas mortality was 12.11 deaths/1,000,000 population. It should be pointed out that the DATASUS website does not reflect the practice of private medicine and that the accuracy of the differential diagnosis of interstitial lung diseases, as well as the completeness of death certificates, is not optimal in Brazil. On the basis of data from the 2010 Brazilian National Census<sup>(5)</sup> and the rates reported in the study cited above,<sup>(4)</sup> 923 new cases of IPF and 2,310 IPF-related deaths were expected in that year. Admittedly, these numbers sound too low, which is likely attributable not only to underreporting but also to a lack of understanding of the disease and to underdiagnosis.

In the absence of data on IPF prevalence in Brazil, we can attempt to calculate the number of affected individuals by speculating on the basis of data available from other countries. Obviously, this approach is imprecise and can lead to conflicting results, depending on the rates adopted.

For that analysis, we chose the rates obtained in two studies conducted in the United States, a country that, like Brazil, has received and still receives a significant influx of immigrants. One of the studies was published in 1994, a time when the definition of IPF was still imperfect.<sup>(6)</sup> However, its methodology was robust, and data were collected in a largely Latin-American population, which, once again, is important for the extrapolation of data for Brazil. The second study was published in 2006, and its strong points include the use of two definitions of IPF, a narrow one and a broad one, as well as the gathering of data from a single large health care plan.<sup>(7)</sup> In addition, in both studies, rates are presented by age group, which is very important for the correction of possible distortions resulting from the different population profiles of the two countries. Finally, the two studies were conducted during a period when, similar to what is currently the case in Brazil, truly effective treatments for the disease were not available. In contrast, the Brazilian population data were obtained from the 2010 National Census.<sup>(5)</sup>

When we applied the rates from the studies conducted in the United States, stratified by age group and gender,

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**Table 1.** Epidemiological data on idiopathic pulmonary fibrosis in Brazil, as calculated on the basis of the rates reported in two studies conducted in the United States and data from the 2010 Brazilian National Census.<sup>(5)</sup>

Calculations based on the broad definition of idiopathic pulmonary fibrosis in Coultas et al. <sup>(6)</sup>		
Age bracket	Annual incidence	Prevalence
>75 years	4,133	6,282
65-74 years	2,881	7,462
55-64 years	1,770	2,364
45-54 years	689	1,843
35-44 years	523	353
<b>Total</b>	<b>9,997</b>	<b>18,305</b>
Calculations based on the narrow definition of idiopathic pulmonary fibrosis in Raghu et al. <sup>(7)</sup>		
Age bracket	Annual incidence	Prevalence
>75 years	1,495	3,540
65-74 years	1,608	3,320
55-64 years	1,623	3,126
45-54 years	1,271	2,430
35-44 years	621	1,103
18-34 years	223	426
<b>Total</b>	<b>6,841</b>	<b>13,945</b>

to the Brazilian population data, we obtained the results listed in Table 1. From Table 1, we might assume that the annual incidence of IPF cases is between 6,841 and 9,997 cases/100,000 population, whereas its prevalence ranges between 13,945 and 18,305 cases/100,000 population. Because IPF is quite rare in young people, if we limit the analysis only to the  $\geq$  55-year age bracket, the projected prevalence might be between 9,986 and 16,109 cases/100,000 population.

From what has been discussed above, we can conclude that, although IPF is a rare disease, it seems to affect a significant number of Brazilians who already require specialized attention and care. With the introduction of the use of new drugs, the survival of patients with

IPF will likely increase and, consequently, so will their care needs.

We point out the equal importance of the fact that speculating on the basis of calculations based on rates from other countries is highly unsatisfactory. Therefore, pulmonologists, epidemiologists, academic institutions, and government bodies, together with patients and their families, should develop initiatives to ensure a better understanding of the epidemiology and natural history of IPF in Brazil. Such initiatives should include not only the creation of databases and registries but also, once those instruments have been developed, the ongoing provision of appropriate information to these systems by medical specialists.

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## ERRATUM

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On page 561 of the original publication, right column, first paragraph, lines two to nine, where it is written

“From Table 1, we might assume that the annual incidence of IPF cases is between 6,841 and 9,997 cases/100,000 population, whereas its prevalence ranges between 13,945 and 18,305 cases/100,000 population. Because IPF is quite rare in young people, if we limit the analysis only to the  $\geq 55$ -year age bracket, the projected prevalence might be between 9,986 and 16,109 cases/100,000 population.”

it should read

“From Table 1, we might assume that the annual incidence of IPF is between 6,841 and 9,997 cases (3.5-5.1/100,000 population), whereas its prevalence ranges between 13,945 and 18,305 cases (7.1-9.4/100,000 population). Because IPF is quite rare in young people, if we limit the analysis only to the  $\geq 55$ -year age bracket, the projected prevalence might be between 9,986 and 16,109 cases (5.1-8.3/100,000 population).”