

Economic Burden of Progressive Pulmonary Fibrosis (PPF) in the Brazilian Private Healthcare System

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Carlos F. S. De Santana<sup>1</sup>, Gabriela Assunção<sup>1</sup>, MD, Magali Caregatti<sup>2</sup>, Robson B. de Oliveira<sup>2</sup>, Taís B. T. Fernandes <sup>2</sup>.  
1 Boehringer Ingelheim Brazil, São Paulo, Brazil; 2 Orizon Healthtech, Barueri, Brazil.

Introduction

Progressive Pulmonary Fibrosis (PPF) is a clinical phenotype observed in patients with fibrosing interstitial lung diseases (ILDs) who experience ongoing progression. It is characterized by worsening respiratory symptoms, declining lung function, and increasing fibrosis on high-resolution computed tomography (HRCT)<sup>1-2</sup>. PPF often develops in patients with underlying autoimmune or connective tissue diseases, such as systemic sclerosis and rheumatoid arthritis, reflecting the heterogeneous and multifactorial nature of fibrotic ILDs<sup>1,3</sup>.

PPF carries a poor prognosis, with a median survival of approximately three to five years after disease progression<sup>4</sup>. Globally, between 13% and 40% of ILD patients evolve to a progressive fibrosing phenotype, with prevalence estimates ranging from 2.2 to 28 cases per 100,000 inhabitants<sup>5</sup>. The disease imposes a substantial healthcare and economic burden, largely due to recurrent diagnostic testing, oxygen therapy, hospitalizations, and prolonged disease management<sup>6</sup>.

Objective

In Brazil, data on PPF remain limited, particularly within the private healthcare system, suggesting underdiagnosis and underreporting., limited monitoring and possible underdiagnosis may obscure the true magnitude of the disease. This study aims to evaluate the direct economic impact and healthcare resource use associated with PPF in Brazilian private healthcare system.

Methods

- A retrospective cohort study was conducted using real-world data from a private administrative claims database, which comprises transactional healthcare data from over 11 million beneficiaries covered by private health plans in Brazil—equivalent to approximately 23% of the national population covered by supplementary health.
- The dataset includes records of medical procedures, hospital admissions, outpatient visits, and billing transactions across a 13-year observation period (January 2010 to December 2023).
- Two distinct patient groups were analyzed:
  - Group 1 comprised individuals with prior events related to underlying ILDs who were subsequently identified with a new event including one of ICD-10 codes of “J84.X” (J84, J84.0, J84.1, J84.8, J84.9). For the purpose of this analysis, these patients were considered to have potentially progressed to PPF following the initial ILD diagnosis. This group was used to reflect longitudinal disease progression.
  - Group 2 included all beneficiaries with at least one recorded ICD-10 “J84.X” event, regardless of prior clinical history, representing the broader population with identified fibrosing pulmonary disease.
- The inclusion of Group 2 was intended to strengthen the reliability and contextualization of the findings by offering a broader epidemiological perspective, in order to assess similarities and differences.
- Healthcare resource utilization was measured based on claims related to chest CT, spirometry (pulmonary function testing), oxygen therapy, and hospital-based care across different modalities of service.
- Total direct medical costs were calculated for each patient over a 12-month period following the first occurrence of J84.x coding.
- Additionally, length of database stay (stay-time) was used as a proxy for longitudinal follow-up. Procedure categories were selected for their clinical relevance in the diagnosis and management of fibrosing ILDs<sup>4,6</sup>.

Results

- A total of 15,526 patients with underlying ILDs were identified in Group 1. Among them, 94 patients (0.6%) where consider progressed to PPF, as defined by a new ICD-10 J84.x event following the base disease diagnosis—yielding an incidence of 605 cases per 100,000 ILD patients. The majority were adults aged 40 to 64 years, with a slight predominance of female sex among those with complete data.

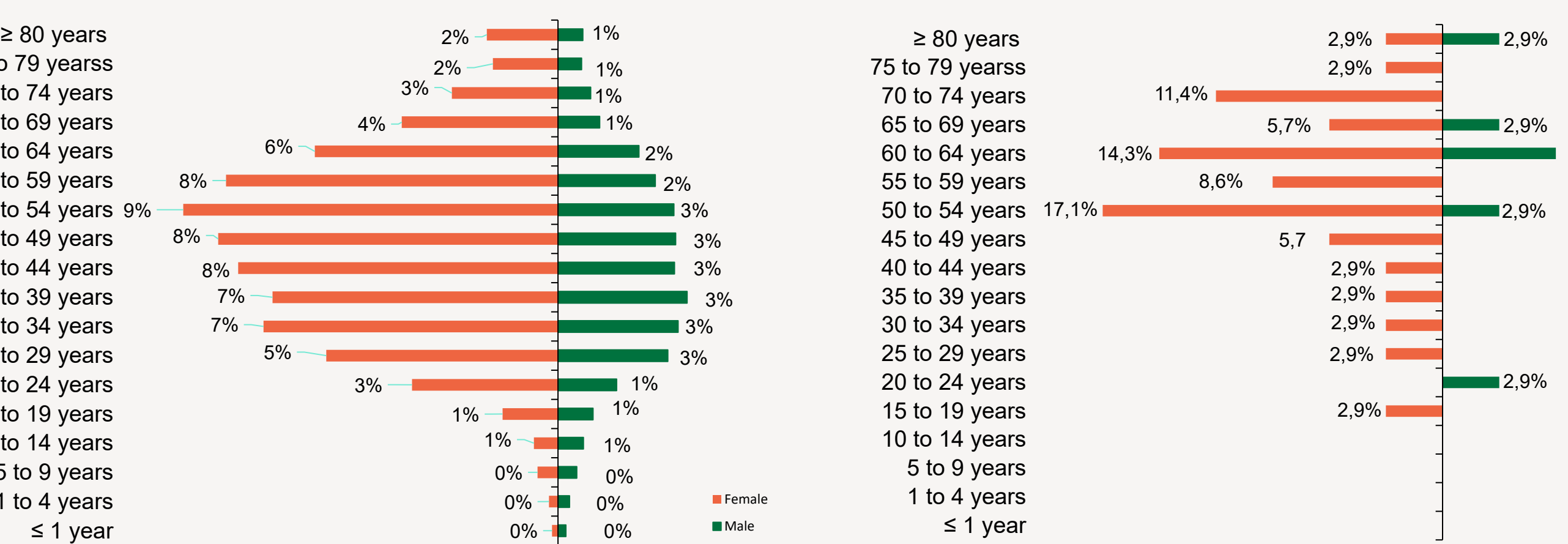


Figure 1. Group 1 age and Sex Distribution among base patients\*  
Figure 2. Group 1 age and Sex Distribution among patients likely to have progressed from ILD to PPF\*

\*Note: Analysis includes only patients with reported age information; patients without recorded age data were excluded from this representation.

References

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- Group 2 included 4,174 patients with at least one J84.X diagnosis between 2012 and 2022, regardless of clinical history. This cohort had a broader age distribution and a higher overall volume of healthcare interactions (figure 3).

Healthcare costs:

- Over the 13-year observation period, the 15,526 patients in Group 1 generated a total healthcare expenditure of BRL 2.82 billion, while the 4,174 patients in Group 2 accounted for BRL 1.17 billion. Among those who progressed to PPF (n = 94), the total expenditure reached BRL 4.95 million, representing approximately 0.15% of the overall costs within Group 1.
- For methodological consistency, all patients were aligned to a common reference point (T0), defined as the first recorded ICD-10 J84.x event. Based on this alignment, healthcare resource utilization and direct medical costs were analyzed within the first 12 months following the index date, leading to the following findings:
  - For the the subgroup of 94 patients from Group 1 who progressed to PPF accounted for approximately BRL 4.8 million in total costs, corresponding to a mean expenditure of BRL 50,728 over the follow-up 12 months period per patient.
  - In contrast for 4,174 patients were identified in Group 2, with a fibrosing event (ICD-10 J84.x), generating cumulative healthcare expenditures of BRL 396.4 million over the same timeframe , with a mean cost of BRL 94,975 per patient.
  - Table 1 summarizes the number of patients in each group who underwent at least one of the selected procedures of interest during the 12-month period, along with the associated total costs.

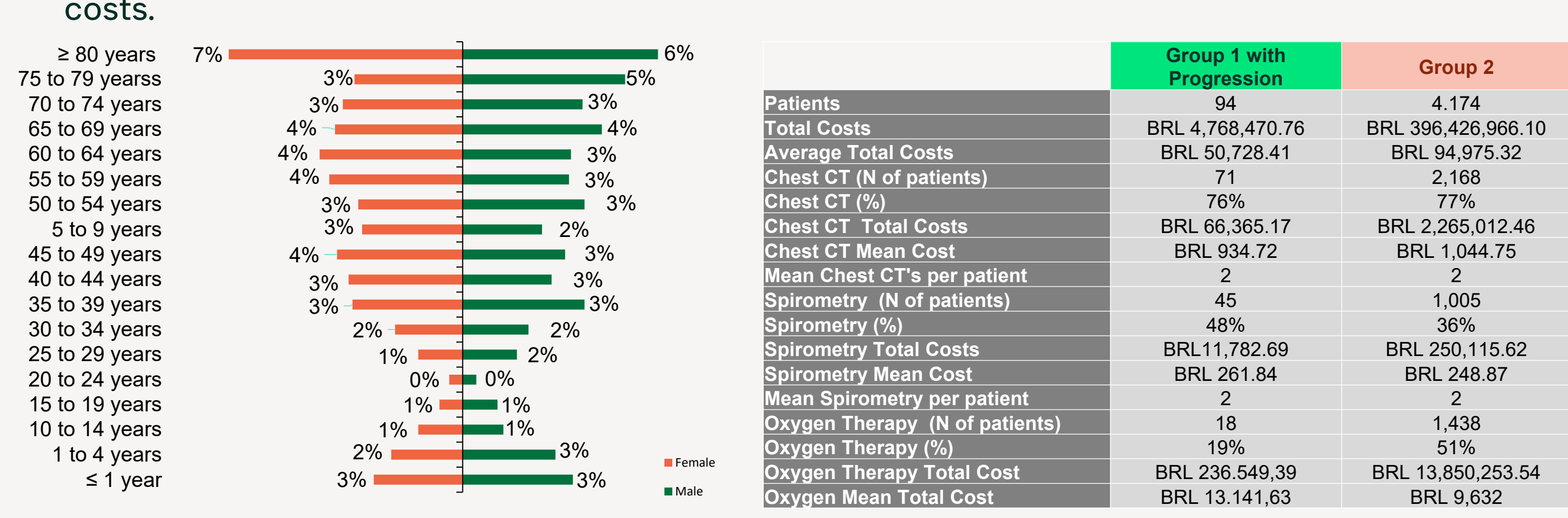


Figure 3. Group 2 age and Sex Distribution among base patients\*  
Table 1. summarizes procedures and total costs per group within the first year after the first event.

Procedures of interest:

- In the Group 1, among patients who progressed, for events related to J84.X directly 89% underwent at least one of the selected procedures of interest during the follow-up period, resulting in a total procedural cost of BRL 234 thousand and a mean of BRL 2,498 per patient. Chest CT was the most frequently performed procedure (76%), followed by spirometry (48%) and oxygen therapy (10%).
- On Group 2, 2,819 patients (67.5%) underwent at least one of the procedures of interest within the 12 months following the first event, resulting in a total procedural cost of BRL 16.4 million (mean: BRL 5,805 per patient). Chest CT was the most frequently performed procedure (77%), followed by spirometry (51%) and oxygen therapy (36%).

Study Limitations

- This study faces inherent limitations related to the complexity of identifying PPF in administrative databases. The absence of a specific ICD-10 code for Progressive Pulmonary Fibrosis, specially since regulatory changes in Brazil since 2015 eliminated the mandatory reporting of ICD codes, further limiting diagnostic traceability over time requires the use of diagnostic proxies, which may compromise the precision of case identification and contribute to underdiagnosis.
- Additional limitations were identified in the assessment of patient demographics, particularly sex and age, as the reporting of these variables was not mandatory in the dataset, limiting subgroup analyses.
- Also, the evaluation of oxygen therapy utilization posed specific methodological challenges due to heterogeneity in billing practices across institutions. In some cases, oxygen was charged per session or event, while in others, it was billed by volume (cubic meters of gas used). This inconsistency may have introduced variability in cost estimates and limited the comparability of oxygen-related expenditures across patients and institutions.
- Together, these limitations highlight systemic barriers to timely PPF identification, earlier clinical suspicion, and structured monitoring protocols to support earlier intervention.

Conclusions

Progressive pulmonary fibrosis (PPF) imposes a substantial clinical and economic burden on Brazil's private healthcare system. Patients who progressed from underlying interstitial lung diseases (Group 1) demonstrated a high intensity of healthcare resource use, despite representing a small fraction of the total cohort. Comparisons with the broader population diagnosed with fibrosing disease (Group 2) revealed important gaps in disease recognition and monitoring.

The findings reinforce the need for earlier identification of disease progression, structured follow-up of high-risk ILD patients, and improved access to diagnostic tools such as high-resolution chest CT. The low rate of imaging and underreporting of ICD codes suggest that many patients remain undiagnosed.

Altogether, these insights highlight clear opportunities to enhance patient care pathways, foster earlier clinical decision-making, and support the integration of innovative therapies for progressive fibrosing diseases in Brazil's healthcare landscape.

Disclosures

- The authors meet criteria for authorship as recommended by the ICMJE.
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