

Cost of Progressive Pulmonary Fibrosis (PPF) in Spain: A Delphi approach

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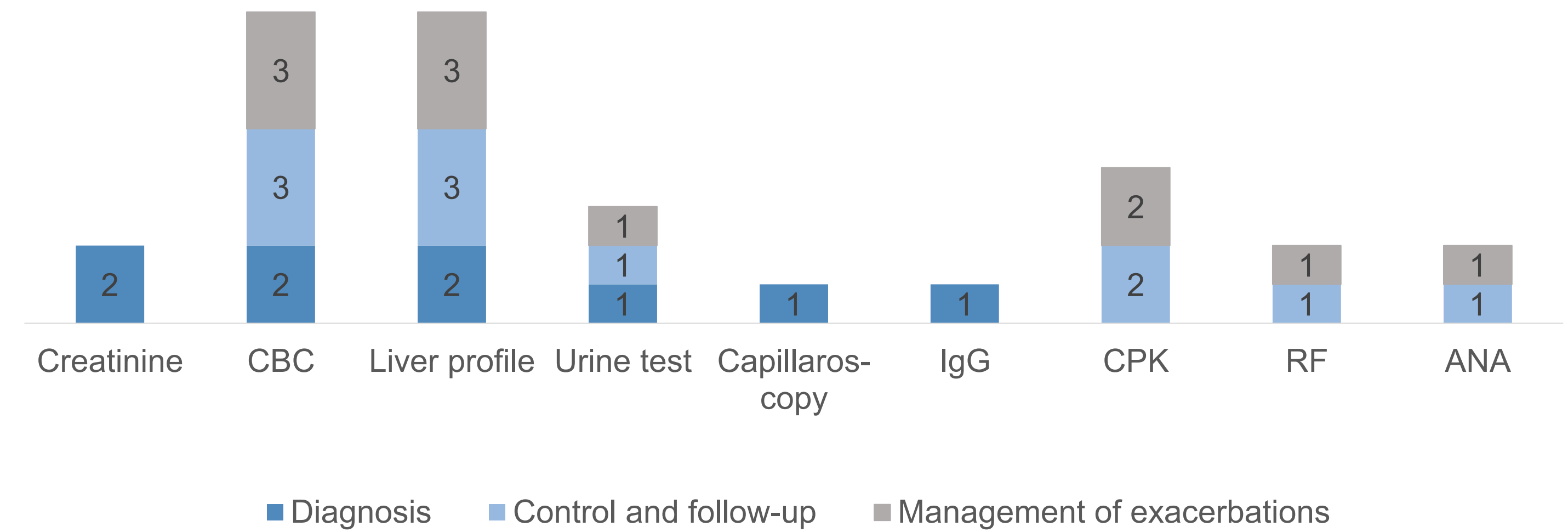
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Introduction

- Interstitial lung disease (ILD) is a heterogeneous group of diseases affecting the alveolar-interstitial structures of the lung, with varying degrees of inflammation and fibrosis¹.
- Some patients with ILD develop a progressive pulmonary fibrosis (PPF), a fatal and limiting disease that persists regardless of the initial trigger of the lung condition²⁻⁵.
- Idiopathic pulmonary fibrosis (IPF) is the best known PF-ILD, with a median survival of 3-5 years without specific treatment^{4,6}. However, progressive fibrosis can occur in other ILD such as, hypersensitivity pneumonitis (HP), fibrotic non-specific idiopathic interstitial pneumonia (NSIP), unclassified ILD, sarcoidosis, mixed connective tissue disease (MCTD), rheumatoid arthritis (RA) and systemic sclerosis, leading to a clinical course similar to IPF, with clinical, functional and/or radiological worsening, respiratory failure and premature death^{2-5,7,8}.
- Considering a prevalence of ILD between 1.73 and 9.76 cases per 10,000 inhabitants^{9,10} and assuming that between 18-32% of patients with non-IPF ILD may develop progressive fibrosis¹¹, it is estimated that PPF affects about 4,700 people in Spain^{12,13}.
- The overall burden of PPF is still unknown. Therefore, this study aims to estimate the use of resources and direct health care cost of PPF in Spain.

- In addition, these patients undergo an average of 2 spirometry evaluations and 1 chest X-ray and High-Resolution Computed Tomography (HRCT) per year.

Figure 2. Median number of laboratory and imaging test per patient



CBC: Complete blood count; IgG: Immunoglobulin G; CPK: Creatine phosphokinase; RF: rheumatoid factor; ANA: antinuclear antibody

Methods

- The Delphi methodology was used to understand the resources needed for the management of PPF in Spain.
- To obtain a better approximation to real clinical practice, in addition to pulmonologists, experts in the management of ILD, to assess the management of autoimmune ILD more broadly, specialists in rheumatology and internal medicine were also included.
- A two-round Delphi panel of 28 experts in autoimmune and non-autoimmune ILD, including pulmonologists (61%), rheumatologists (21%) and internal medicine (18%), was carried out between January and December 2020.
- The resources evaluated were the median number of medical visits, laboratory tests and imaging tests performed: at diagnosis, during disease control and follow-up, and for the management of exacerbations.
- The direct costs of the disease, expressed as average and interquartile range, were estimated from the identified median number of each resource and the corresponding unit cost, obtained from the eSalud Spanish database.

Hospitalizations

- During the course of the disease, on average, these patients require an annual hospital admission of 8 days in the pulmonology department. Additionally, due to exacerbation episodes, they also require an annual hospitalization of 11 days in the pulmonology department and an ICU admission of 9 days, per year (Table 1).

Table 1. Median number and duration of hospitalizations per year

	Control and follow-up		Exacerbations	
	Number of hospitalizations	Average length of stay (days)	Number of hospitalizations	Average length of stay (days)
Pulmonology department	1	8	1	11
Intensive care unit	-	-	1	9

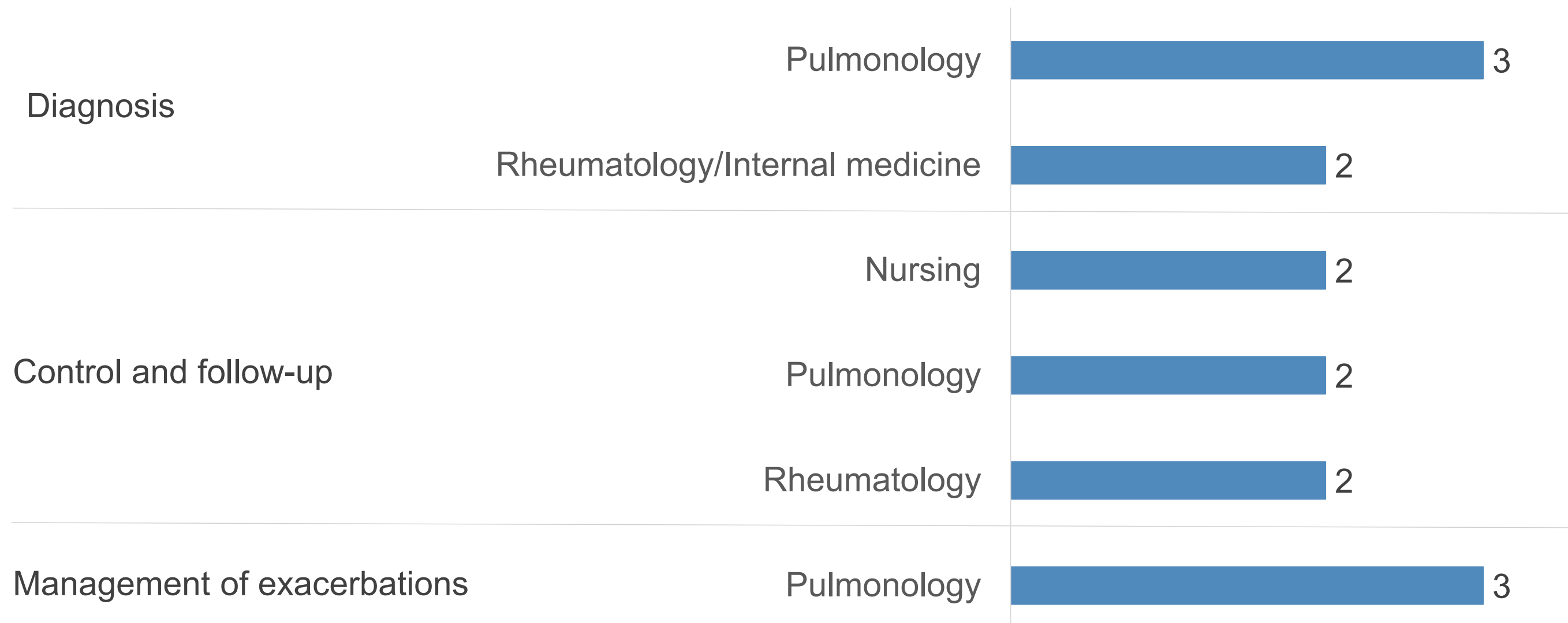
Results

USE OF RESOURCES

Medical visits

- During the year prior to the definitive diagnosis of PPF, the experts agreed that, at least, an average of 3 visits to the pulmonologist and 2 visits to the rheumatologist/internal medicine are necessary. Additionally, during the disease control and follow-up an average of 2 visits to pulmonology, rheumatology and nursing are required, as well as an average of 3 visits to pulmonology for the management of an acute exacerbation (Figure 1).

Figure 1. Median number of medical visits per patient



Laboratory and imaging tests

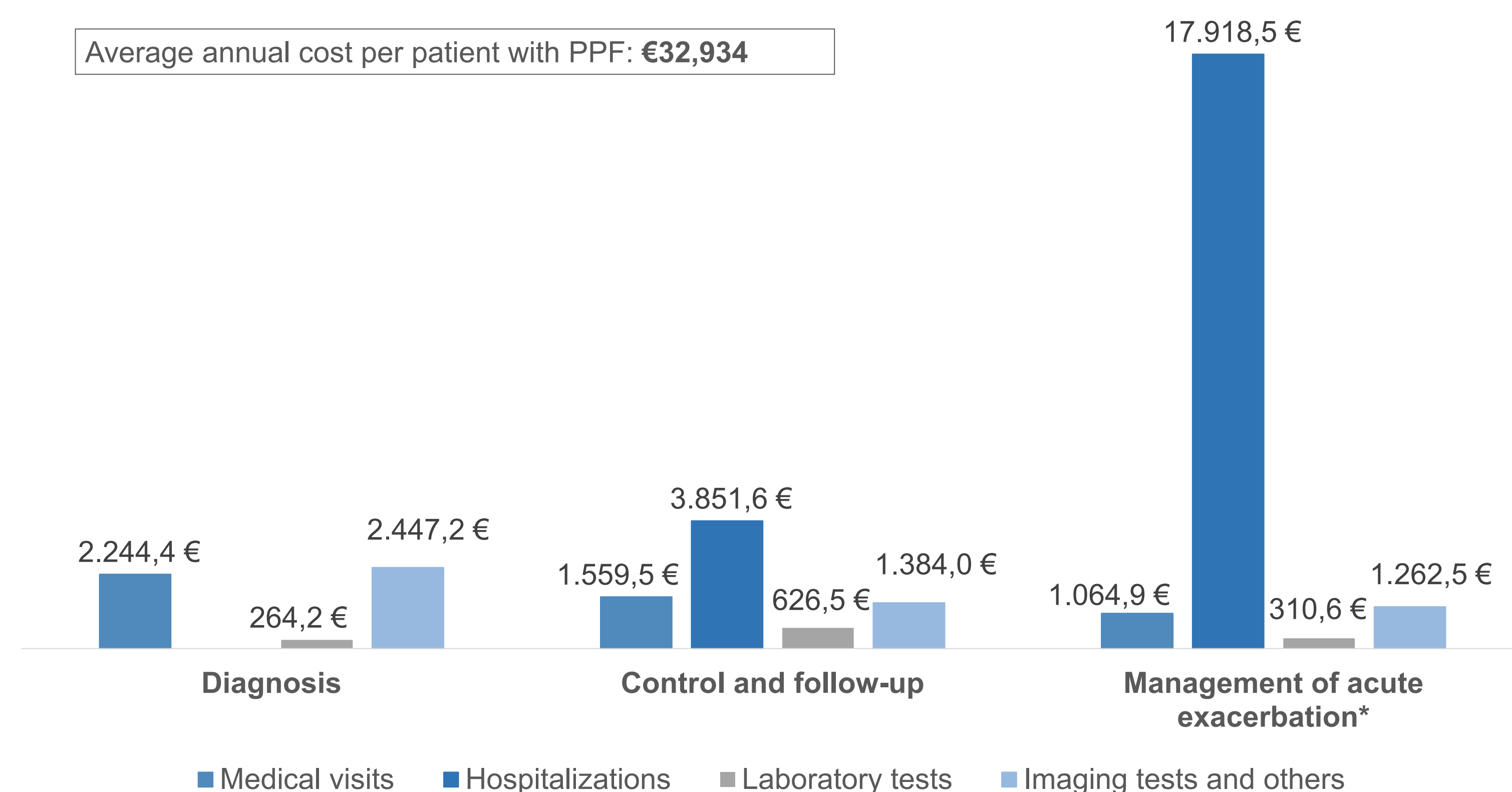
- According to experts' opinion, the most common laboratory tests carried out during the course of the disease are, complete blood count, urine test and liver profile, performed once to twice a year during diagnosis and up to 3 times a year during the follow-up and management of acute exacerbation (Figure 2).

COSTS

Average health care cost per patient with non-IPF PPF

- As a result, the estimated cost associated to diagnosis, control and follow-up and management of acute exacerbation was €4,955.9, €7,421.6 and €20,556.5, respectively, leading to an annual total cost of €32,934.0 per patient with PPF (Figure 3).
- The annual cost per patient with PPF was similar to the that estimated by Morell et al. in patients with IPF (€26,435 per year)¹⁴.
- Management of acute exacerbations was the biggest cost driver of the disease, mainly due to the high cost associated to ICU admissions (€17,918.5).
- These costs are estimated and have been obtained by considering those values where there was a consensus among experts and those where there was not and, therefore, the validity of these may be relatively low.

Figure 3. Average annual cost per patient with PPF



*According to pulmonologists, patients with PPF suffer around 1 exacerbations/year. PPF: progressive pulmonary fibrosis

Conclusions

- These results highlight the high economic impact associated to the disease, mainly due to the cost of ICU admission during the management of acute exacerbations.
- In accordance with the estimated cost in patients with IPF (€26,435 per year)¹⁴, the average cost per patient with non-IPF PPF amounts to €32,933.96 per year.

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