

## Awareness towards the main ILD among primary care physicians

Mariana Silva,<sup>1\*</sup> Amélia Fernandes,<sup>2</sup> Ana Rita Pereira,<sup>3</sup> Sofia Madanelo,<sup>4</sup> Tatiana Clemêncio,<sup>5</sup> Pedro Gonçalo Ferreira<sup>6\*</sup>

*\*These authors contributed equally.*

<sup>1</sup>Unidade de Cuidados de Saúde Personalizados Anadia II, Anadia

<sup>2</sup>Unidade de Saúde Familiar Vale do Cértima, Oliveira do Bairro

<sup>3</sup>Unidade de Cuidados de Saúde Personalizados Anadia I, Anadia

<sup>4</sup>Unidade de Saúde Familiar Flor de Sal, Aveiro

<sup>5</sup>Unidade de Saúde Familiar de Santa Joana, Cacia

<sup>6</sup>Serviço de Pneumologia, Centro Hospitalar e Universitário de Coimbra, Portugal

### ABSTRACT

**Background:** Interstitial lung diseases (ILDs) comprise a group of multiple entities sharing some clinical, functional, and radiological similarities. In many countries primary care setting has been devoid of pre- and post-graduate educational interventions focused on basic knowledge on ILD. This, along with usual nonspecificity of symptoms at presentation, may contribute to diagnostic delay in this disease setting.

**Methods:** We designed a study questionnaire to assess the level of awareness on basic diagnostic and management aspects of core ILDs – idiopathic pulmonary fibrosis (IPF), hypersensitivity pneumonitis, sarcoidosis, connective tissue disease related-ILD, and drug-induced ILD - among primary care physicians (GPs) from five “ACeS Baixo Vouga” health centres and to perceive possible weaknesses. Differences in awareness between GPs under 45 and over 45 years-old were also assessed.

**Results:** Globally, 69% of questions were correctly answered but only 21.9% of GPs considered to have a satisfactory self-perceived level of knowledge on ILD. Except sarcoidosis ( $p=0.017$ ) and some isolated questions on other diseases, no significant differences were found between physicians below 45 years and above. Though, there was a trend to higher awareness in the younger group. The best awareness was seen in sarcoidosis. IPF questions had the worst performance and only 48.5% of GPs recognized the importance of velcro-type crackles in suggesting a possible diagnosis.

**Conclusion:** Specific attention should be devoted to educational interventions directed to GPs on basic notions on the main ILDs. This could improve the usual diagnostic delay in many ILDs, as a timely diagnosis is essential for an early treatment and prolonged patient survival.

**Key words:** Interstitial lung diseases; primary care.

**Correspondence:** Pedro Gonçalo Ferreira, Serviço de Pneumologia, Centro Hospitalar e Universitário de Coimbra, Praceta Mota Pinto, 3004-561 Coimbra, Portugal. E-mail: pedrogferreira8@gmail.com

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## Introduction

Interstitial lung diseases (ILDs) comprise a group of multiple entities with different relative rarity and often sharing some degree of similarities, regarding their clinical, functional, and radiological manifestations. Many of these diseases, particularly progressive fibrosing ILDs, usually inflict substantial burden to patients, their families, and to the healthcare systems. Among the most prevalent entities one can find: sarcoidosis, idiopathic pulmonary fibrosis (IPF), hypersensitivity pneumonitis (HP), and connective tissue disease related-ILD (CTD-ILD). Although ILDs are classically managed by dedicated pulmonologists, primary care physicians (GPs) are often at the interface with symptomatic patients and, accordingly to the prerogative of family medicine, they tend to follow these same patients and their families longitudinally. Desirably, an interconnected dialogue between the two medical specialties during both pre-diagnostic assessment and patient follow up could be argued as a quality standard. Nonetheless, in many countries primary care setting has been chronically devoid of pre- and post-graduate educational interventions attentive on basic knowledge about ILD, namely non-existing contact with dedicated ILD clinics and multidisciplinary ILD meetings within the postgraduate internships in pulmonology departments, also not including ILD in the context of respiratory update courses (chronically devoted to COPD and asthma).

Importantly, aspects such as the semiological recognition of “velcro” like-rales, knowledge of the demographic profile of some diseases, the proper valorisation of certain inhalational exposures (organic antigens and occupational chemicals) or drugs, and knowing the potential pulmonary manifestations of certain connective-tissue-diseases, may all be decisive to an early diagnostic insight. The non-acknowledgment of this clues, along with the usual nonspecificity of symptoms at presentation, can be contributing causes for the diagnostic delay oftenly found in various forms of ILD. Namely, in IPF it has been shown that patients commonly experience significant delays in receiving an accurate diagnosis, with recent series showing a mean delay of about 2.1 years from symptom onset to diagnosis [1,2]. This can lead to a delayed start of effective treatment and a negative influence on the disease course and prognosis [1,2].

Also, given the relatively recent advances on the pathobiological understanding [3] and diagnostic [4] and therapeutic management [5] of some of these entities and the growing role of ILD in the scope of many clinical journals, different awareness between recent and older generations of primary care physicians could possibly exist.

Therefore, we designed a study to assess the level of awareness regarding general aspects of ILD diagnosis and management of core ILDs - IPF, HP, sarcoidosis, CTD-ILD and drug-induced ILDs

- among primary care physicians working on the five health care centres of “ACeS Baixo Vouga”, located in the central region of Portugal. These five community health care centres usually refer respiratory patients to the Pulmonology Department of Centro Hospitalar do Baixo Vouga, where a dedicated ILD outpatient clinic and multidisciplinary ILD team were place, receiving around 160 new patients yearly.

## Methods

This was an observational cross-sectional study by application of a multiple-choice questionnaire (Appendix A) with 38 questions, under anonymity, to all the primary care doctors (specialists and residents) from five AceS Baixo Vouga health units. These five health units follow around 58,000 patients.

In the first part of the survey, demographic characteristics of physicians (age, gender) were assessed and used to demarcate two age subgroups – “doctors under 45 years old” and “doctors equal or above 45 years old” – for comparative analysis. The second section consisted in a framework of questions regarding general aspects of ILD definition, heterogeneity, “anamnesic pearls” and general diagnostic framework, followed by other subgroups of intermixed questions related to basic diagnostic profiles and management features of IPF, HP, CTD-ILD, sarcoidosis and drug-induced ILD. Table 1 represents this survey’s questions divided by subgroups, evaluated by our analysis.

The results were used to assess the level of knowledge on ILDs in the setting of primary care practice, perceive possible specific weaknesses and eventually reflect on possible solutions to improve the level of proficiency related to some basic concepts. Differences in awareness between <45 years and ≥45 years doctors were also assessed. Data was recorded with the Excel 2010 and SPSS 22. Categorical variables were reported as number (%). Levene’s test for independent samples were used to assess differences by age group. A p lower than 0.05 was considered to be statistically significant.

## Results and discussion

The mean age of the responding physicians was approximately 43 years (SD ± 14.9), with a clear female predominance (81.8%), otherwise reflective of the allocated human resources currently working in the 5 targeted centres. The younger (<45-years-old) and the older subgroup (≥45-years-old) comprised 18 (54.5%) and 13 (39.4%) physicians, respectively. Two doctors (6.1%) have not informed about their age. Globally, 69% (832 of the 1,205) of the questions were correctly answered. Among the remaining 31%, 73% were wrong answers and 27% were “do not know” answers.

No statistically significant differences were found between the

**Table 1. Subgroups of questions.**

	Questions
I Concept, semiology and general management of ILD	2, 4, 5, 6, 8, 10, 12, 16, 17, 18, 19, 22, 23, 25, 27, 28, 31, 32, 34, 36, 38
II Features conducive to early diagnostic suspicion	5, 9, 12, 13, 16, 17, 27, 28, 34, 36
III Idiopathic pulmonary fibrosis (IPF)	5, 7, 8, 12, 14, 15, 17, 19, 20, 21, 24, 26, 31, 35, 37
IV Sarcoidosis	9, 29, 33
V Hypersensitivity pneumonitis (HP)	3, 11, 21, 27, 30, 31, 34
VI Connective tissue disease interstitial lung disease (CTD-ILD) and drug induced-ILD	4, 13, 28, 31

subgroup of physicians below 45 years of age (younger subgroup) and those aged  $\geq 45$  (older subgroup), except for the sarcoidosis group ( $p=0.017$ ) (Table 2) and some specific isolated questions:

- Concept, semiology and general management of ILD group: question 18 ( $p=0.009$ ) with better results in the older GPs subgroup ( $\geq 45$  yrs), and questions 23 ( $p=0.006$ ), 25 ( $p=0.001$ ), 32 ( $p=0.011$ ) and 38 ( $p=0.000$ ) with better results in the younger GPs subgroup ( $<45$  yrs);
- IPF group: question 7 ( $p=0.012$ ) with better results in the younger GPs subgroup;
- Sarcoidosis group: The overall result to this subgroup of questions was statistically better in the older subgroup ( $p=0.017$ );
- HP group: questions 3 ( $p=0.000$ ) and 30 ( $p=0.006$ ), both with better results in the younger GPs subgroup;
- CTD-ILD and drug induced-ILD group: question 13 ( $p=0.000$ ) with better results in the younger GPs subgroup.

Concerning the first question of the questionnaire – “As a GP, I feel that I have sufficient knowledge about ILD for what my duties are” - we observed that only 21.9% of the global GPs considered having, in fact, a satisfactory self-perceived level of knowledge on ILDs. In the subgroup  $<45$ -years-old, an affirmative answer was given by 27.8% and in the subgroup of physicians  $\geq 45$ -years-old, remarkably only 8.3% were comfortable with their self-perceived level of knowledge. Possible reasons for this difference may have been the fact that some of the younger physicians of the five health care centres had previously taken contact with the hospital ILD outpatient clinic in the setting of their programmed 2-month (optional) internship on pulmonology. Nonetheless, 69% of the total questions were correctly answered, which was quite better than what could be expected based on their self-perception of knowledge.

Regarding basic features and concept of ILDs, awareness to key semiology findings and general diagnostic steps, 61.2% of these subsets of questions were correctly answered by the group, with 22.2% choosing a wrong answer and 16.6% choosing “do not know” (Table 2). Nonetheless, no GP achieved all the correct answers on this matter and, worryingly, only 48.5% and 60.6% of the total GPs recognized the importance of finding velcro-crackles and digital clubbing, respectively, to elicit the possibility of a fibrosing ILD [6].

A better result was achieved in acknowledging the value of a proper (high-resolution) chest CT protocol to investigate a possible ILD, with 78.8% of correct answers. Importantly, the possible presence of relevant weight loss and anorexia was associated only to lung cancer and not presumed possible in ILD by 21.2% of the

GPs. Presently there is solid evidence that decrements in body mass are predictive of increased mortality in ILD, specially IPF [7,8].

Taken together, the roll of questions encompassing an early insight to the possibility of ILD (Table 1) and the chance to a timely referral to a hospital ILD outpatient clinic had 70.7% of global correct answers, though only 9.1% of the responding physicians had a completely correct set of answers. The older GPs subgroup was found to have a lower performance (64.8% versus 73.9% in the younger subgroup) (Table 2). The nosological diagnosis associated with the best awareness was sarcoidosis, with 94.9% of correct answers (Table 2). Sarcoidosis was easily associated to both a tendentially younger age of incidence and to the possibility of acutely presenting itself as Lofgren syndrome (questions 9 and 33). The possibility of extrapulmonary involvement was acknowledged by 96.9%, with better results in the older GPs subgroup. This result may possibly relate to the higher prevalence of this disease among ILDs and, thus, to a greater representativeness in the patient files of each GP.

The IPF set of questions had the worst performance with only 55.9% of correct answers, with a lower performance by the GPs  $\geq 45$  yrs (50.0% of correctly answered questions) (Table 2). Clearly there was unawareness regarding the importance of forced vital capacity to monitor IPF patients (question 7), with only 38.7% of correct answers, possibly reflecting a rigid focus on forced expiratory volume in the first second ( $FEV_1$ ) due to educational interventions traditionally focused on COPD and asthma. Another weak point was related to the concept, indication and purpose of antifibrotic agents in IPF and the risk associated to steroids (questions 19 and 20 with 9.1% and 3% correct answers, respectively). These results could be explained by the considerable progress made in recent years in the pathophysiological understanding of the disease, with a departure from the traditional model of post-immunoinflammatory damage to the current concept of primarily fibrotic disease. The unawareness regarding the positioning of pirfenidone and nintedanib can be framed in the fact that treatment management falls entirely on hospital clinics and on the absence of educational initiatives on IPF focused on primary care. Another interesting aspect regards to the impact of specific comorbidities in IPF. Globally about 30.3% of the GPs showed not to be aware of the potential impact of untreated obstructive sleep apnea [9] or symptomatic gastroesophageal reflux [10,11].

The set of questions addressing awareness towards HP was successfully answered, with 82.8% of globally correct answers with a superior performance of the younger GPs subgroup (87.3%

**Table 2. Performance by subgroup of questions and age group.**

	Total group (correct answers %)	Global performance <45 yrs physicians (correct answers %)	$\geq 45$ yrs physicians (correct answers %)	p*
Concept, semiology and general management of ILD	61.2%	63.2%	55.1%	0.329
Early disease suspicion	70.7%	73.9%	64.8%	0.177
Idiopathic pulmonary fibrosis	55.9%	59.5%	50.0%	0.265
Sarcoidosis	94.9%	92.4%	97.4%	0.017*
Hypersensitivity pneumonitis	82.8%	87.3%	74.7%	0.903
CTD-ILD and drug induced-ILD	81.5%	83.3%	76.0%	0.322

\*t-test for independent; ILD, interstitial lung disease; CTD-ILD, connective tissue disease-related ILD.

vs 74.7% in the older subgroup) (Table 2). Globally, 87.9% of the GPs considered to be familiar with the designation of the disease, 7.8% knew that HP is a disease with variable prognosis and 87.9% understood how important the avoidance of specific antigen exposures can be for treatment and prognostic purposes. Although, the fact that 12.1% did not address the possibility of isocyanate induced-HP can easily point to the risk of insufficient insight for the diagnostic possibility in certain settings.

The performance related to CTD-ILD and drug induced-ILD also presented good results, with 81.5% of correct answers, once again with better outcomes in the younger GPs subgroup (83.3% vs 76.0% correct answers) (Table 2). Namely, 78.8% of GPs recognized the fact that interstitial lung disease may be a privileged manifestation of systemic sclerosis and rheumatoid arthritis [12]. However, importantly nearly one third of the GPs (34.4%) were not aware that nitrofurantoin and amiodarone are among the drugs more commonly associated with drug-induced ILD (question 28) [13,14].

Respiratory rehabilitation is presently positioned as an important strategy in the treatment of ILD patients, able to provide improvements in exercise capacity, dyspnoea and quality of life, at least in the short term [15,16]. The potential applicability of respiratory rehabilitation was also addressed in the questionnaire with 90.9% of physicians acknowledging that this type of intervention can also have a role in the ILD setting.

Lastly, regarding patient follow up 93.9% of primary care physicians think ILD patients should have a complementary surveillance by both pulmonology and primary care during their longitudinal follow up.

## Conclusions

Regarding their professional role as family doctors and the importance they can sustain in the initial assessment and timely referral of ILD patients, only a minority of GPs were confident on their self-perceived level of awareness.

A pleasing performance was found in the sets of questions relating to sarcoidosis, hypersensitivity pneumonitis, CTD-ILD and drug induced-ILD. On the other hand, a poor level of awareness was evident regarding IPF and on key semiology features and general diagnostic conduct. Globally, GPs  $\geq 45$ -years-old showed weaker confidence and, tendentially, lower awareness compared to the younger generation of colleagues.

According to the GP's perspective, longitudinal follow up of ILD patients should be done in a shared and discussed manner with pulmonology. In this regard, although ILDs have a lower incidence compared to other chronic respiratory diseases, we believe that a more specific attention should be devoted to educational interventions directed to primary care physicians on basic notions on the main ILDs, specially IPF. These may allow the opportune modification of identifiable risk exposures, raising the level of disease-suspicion in the face of certain demographic profiles, semiology findings and reliable systemic diseases, thereby helping to mitigate the diagnostic delay in many ILDs. Indeed, a timely diagnosis is crucial to initiate early treatment, delay disease progression and prolong survival of these patients. Nonetheless, the crucial role of GPs in the early diagnosis demands attention for prior adequate training and preventive educational interventions. These could foster "green corridors" for rapid referral of patients and also facilitate the desirable dialogue between pulmonologists and primary care physicians during patient follow up.

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## Abbreviations

COPD:	Chronic obstructive pulmonary disease;
CT:	Computed tomography;
CTD-ILD:	Connective tissue disease related-interstitial lung disease;
FEV <sub>1</sub> :	Forced expiratory volume in the first second;
GPs:	Primary care physicians;
HP:	Hypersensitivity pneumonitis;
ILD:	Interstitial lung diseases;
IPF:	Idiopathic pulmonary fibrosis.

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