Respiratory Medicine (2009) **103**, 1122–1129

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journal homepage: www.elsevier.com/locate/rmed

Epidemiology of interstitial lung diseases in Greece ☆

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Received 14 August 2008; accepted 2 March 2009
Available online 3 April 2009

KEYWORDS
Registry;
Interstitial lung diseases;
Epidemiology;
Greece;
Idiopathic interstitial pneumonias;
Idiopathic pulmonary fibrosis

Summary
Introduction: Few data are available on the epidemiology of interstitial lung diseases (ILDs), especially after the current classification of idiopathic interstitial pneumonias. The aim of this study is to provide data on the epidemiology of ILDs in Greece, under the ATS/ERS international consensus.
Methods: Departments of Pneumonology were contacted and asked to complete a questionnaire for every case of ILD that was alive on 2004 as well as for every new case from 1st January 2004 to 31st December 2004. Questions on the patients' demographic data, the exact diagnosis and the procedures used to establish the diagnosis were included. Centers covering about 60% of the Greek population have been analyzed.
Results: A total of 967 cases have been registered. The estimated prevalence of ILDs is 17.3 cases per 100,000 inhabitants. The estimated annual incidence of ILDs is 4.63 new cases per 100,000 inhabitants. The most frequent disease is sarcoidosis (34.1%), followed in decreasing order by idiopathic pulmonary fibrosis (19.5%), ILD associated with collagen vascular diseases (12.4%), cryptogenic organizing pneumonia (5.3%), histiocytosis (3.8%), and hypersensitivity pneumonitis (2.6%). Unclassified ILD or not otherwise specified accounted for the 8.5% of prevalent cases.

☆ Presented in part at the 15th Annual ERS Congress, Copenhagen 2005.

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0954-6111/$ - see front matter
doi:10.1016/j.rmed.2009.03.001

Conclusions: These data suggest that sarcoidosis and idiopathic pulmonary fibrosis are the most frequent ILDs in our population. In comparison with the few previous reports, interesting dissimilarities have been observed.

Introduction

More than 200 different entities are included under the term interstitial lung diseases (ILDs). In their pathogenesis many factors such as environmental and occupational agents, infections, drugs, radiation and genetic predisposition have been implicated.[1–4] However, the majority of these diseases are considered idiopathic, without a curable treatment.[5] Several attempts to estimate the incidence and prevalence of ILDs, although limited by variable and confusing diagnostic criteria, suggest that there is a rising trend in rates worldwide as well as international differences in the prevalence rates of the various forms of ILDs.[6–12] The recent American Thoracic Society/European Respiratory Society [ATS/ERS] international consensus classification of the idiopathic interstitial pneumonias, offers an opportunity for greater precision in epidemiological studies.[1–3]

There are few epidemiologic studies available regarding the incidence, prevalence or relative frequency of ILDs.[6–18] To date, a number of ILD registries have been established in various regions. Apart from the case of New Mexico,[6] the other registries have been organized by pneumonologists, and clearly underestimate the real incidence and prevalence of ILDs.[9–18] However, knowledge of the relative frequency of the diagnosis of the different types of ILD may provide interesting information about these diseases. Furthermore, histopathologic subsets of idiopathic interstitial pneumonias (IIPs) have prognostic significance.[19–21]

The aim of this study is to estimate the prevalence and incidence rate of ILDs in Greece in the light of the new international consensus classification of the IIPs and to compare our findings with those of other reports.

Materials and methods

By the end of 2003 the Interstitial Lung diseases Group of the Hellenic Thoracic Society (HILD) contacted the departments of pneumonology with special interest in ILDs from all over Greece and asked pneumonologists to complete a questionnaire for every case of ILD that was alive on 2004 (prevalent case) as well as for every new case (incident case) diagnosed from 1st January 2004 to 31st December 2004.

In order to achieve a good response rate the questionnaire was kept as simple as possible (one-page, Appendix A). Questions on the patients' demographic data, the exact diagnosis-type of ILD and the procedures, such as high-resolution computed tomography (HRCT), bronchoalveolar lavage (BAL), transbronchial lung biopsy, surgical lung biopsy, and serology, used to establish the diagnosis were included.

In every participating center the expert pneumonologist on ILDs was responsible to identify the cases based on the actual histological and radiological reports from pathology/BAL specimens and HRCT (evaluated by an expert pathologist and an expert radiologist respectively). Due to funding limitations it was not possible to create a panel of experts to visit the different hospitals and review each one of the 967 cases. The working group holds four meetings in order to standardize the diagnostic criteria. For every test performed in each one of the participating hospitals there are internal quality control procedures. In addition, guidelines for classification were repetitively discussed during specific sessions organized by the HILD Group. For the classification of idiopathic interstitial pneumonias' the ATS/ERS international multidisciplinary consensus[1] was used. As for sarcoidosis the diagnostic approaches proposed in the joint statement of the ATS, ERS and the World Association of Sarcoidosis and Other Granulomatous Disorders (WASOG) were applied.[22] ILDs due to neoplastic diseases, infections or heart diseases were excluded from the study.

Centers covering about 60% of the Greek adult population (age greater than 15 years), that is 5.6×10^6 inhabitants, have replied and participated in the study.

The diagnostic process for ILD was the same in all the centers contacted and the diagnostic algorithm used was that proposed by the ATS/ERS international multidisciplinary consensus.[1] Thus, the centers that participated in the study are probably representative of all the centers contacted. No major differences in case ascertainment have been observed among centers.

The study was approved by the Institutional Review Board of Demokritus University of Thrace.

Results

A total of 967 cases have been reported with a small preponderance of female population (53.6%). The male to female ratio was 1:1.15. Mean [SE] age of male patients was 58 [0.82] years and that of female 59.3 [0.64] years. Incidence rate is estimated to be 4.63 per 100,000/year and prevalence 17.3 per 100,000.

Table 1 shows the distribution of prevalent ILD cases and their prevalence in our population. The most frequent disease entity is sarcoidosis (34.1%) followed by IIPs (29.5%). Idiopathic pulmonary fibrosis (IPF) accounted for about 20% of all ILDs, whilst usual interstitial pneumonia (UIP) was the most frequent histology among IIPs. The above-mentioned diseases are followed in decreasing order by ILD-associated collagen vascular diseases (12.4%), cryptogenic organizing pneumonia (5.3%), histiocytosis (3.8%), and hypersensitivity pneumonitis (2.6%). Unclassified or not otherwise specified ILDs accounted for the 8.5% of prevalent cases.

The incidence of ILDs in our population and the number (%) of incident cases are shown in Table 2. Among incident cases, IIPs are the most frequent (32.4%) followed by sarcoidosis (23.2%).