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Patient experiences with pulmonary fibrosis

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Summary

This survey describes the experiences of patients diagnosed with pulmonary fibrosis, focusing on the issues of patient education and resources. A survey of 52 defined-choice and open-ended questions regarding the diagnosis and management of pulmonary fibrosis was delivered. A total of 1448 respondents comprised the study group. Two-thirds of respondents reported a clear lack of information and resources on pulmonary fibrosis at the time of diagnosis. Less than half of respondents reported they felt well-informed about treatment options, the role of supplemental oxygen, pulmonary rehabilitation, and transplantation. These results suggest there is a substantial need for improved patient education regarding the diagnosis and management of pulmonary fibrosis.

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Introduction

Few diagnoses in pulmonary medicine generate as much concern among both patients and their physicians as

pulmonary fibrosis. An accurate diagnosis is challenging as it requires the skilled integration of clinical, radiographic and histopathologic findings that are rarely textbook in presentation.¹ The most common cause of pulmonary fibrosis, idiopathic pulmonary fibrosis pulmonary fibrosis is a devastating diagnosis, as the disease has no effective therapy and its prognosis is uniquely poor.^{2–7}

Significant advances have been made in the diagnosis of pulmonary fibrosis, in particular IPF, in the last decade.

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This includes the definition of IPF as pulmonary fibrosis characterized by usual interstitial pneumonia pattern on surgical lung biopsy,² the demonstration that many patients with IPF can be accurately diagnosed without a surgical lung biopsy in the right clinical context,^{8,9} and the establishment of a clearer understanding of prognosis and predictors of survival.^{5-7,10-12} It is unclear how these advances have impacted community practice, and whether clinicians are translating this knowledge to their patients with pulmonary fibrosis.

The coalition for pulmonary fibrosis (CPF), a non-profit public benefit organization, developed a research questionnaire to collect information from its members regarding patient experiences with the management of IPF and other forms of pulmonary fibrosis. A wide range of topics was covered, with a major focus on the attitudes and experiences of patients diagnosed with pulmonary fibrosis with regards to diagnosis and management of their disease. This paper summarizes the results of this sample survey.

Methods

The CPF is a nonprofit organization founded to further patient and physician support and education, and to enhance research efforts in pulmonary fibrosis. The CPF is governed by pulmonologists, patients, investigators and advocacy organizations. Membership is free, and most members learn of the CPF through seminars, word of mouth or the Internet.

A patient questionnaire was developed in conjunction with clinicians experienced in issues surrounding the management of pulmonary fibrosis in order to better understand the experiences of patients with this condition. The questionnaire was designed to collect the following information: demographics, family history, potential environmental exposures, diagnostic testing performed, treatments prescribed, and patient attitudes regarding the adequacy of information received from health care providers concerning the management of their disease. Questions were created by the authors based on informal interviews with patients and family members of patients including those participating in CPF-sponsored support groups. Draft questions were reviewed by patients and families for clarity, and edits were made by two authors (MM, BC) according to industry standards.

Funding for this project was provided by the DuBrul Family Fund and the family of Helen and Michael Galvin. The questionnaire was distributed to over 2000 members (all patients and caregivers of current and deceased patients for which addresses were available) between July 2003 and December 2004. The questionnaire was sent both in paper form and via email with a link to a web-based survey site. The survey was also made available to CPF members online and in paper form through the CPF website, sponsored seminars and institutional partners.

Completed questionnaires were compiled by Michaels Opinion Research, Inc. (MM, BC), an independent research firm experienced in conducting sample surveys. Results reported in this publication are based on the responses from living patients or caregivers of living patients. Two authors (HRC, MIS) reviewed the raw data and directed the analysis.

Results were tabulated as a whole as well as by age and year of diagnosis. Statistical analysis was performed using WinCross cross-tabulation software. Comparisons across groups (e.g. by year of diagnosis) were made using independent *t*-tests for means and Z-tests for independent proportions. Full results of the survey are published in the electronic appendix to this publication. This project was approved by the University of California San Francisco Committee on Human Research.

Results

Demographics

Responses were received from 1583 individuals. The majority of respondents were current patients (79.0%), the minority were caregivers of current (12.4%) or deceased patients (8.5%). There were 1448 current patients or caregivers of current patients who responded (current patients ($n = 1251$), caregivers responding on behalf of current patients ($n = 197$)), and these make up the study cohort. Responses were generally similar between patients and caregivers and are reported together throughout this publication.

The median age of respondents was 65 years (Table 1). There were slightly more men than women (1.2:1), and the vast majority were white (90.2%). Sixty-nine percent (68.9%) reported a history of cigarette use (mean 24.5 years), including only 3.2% currently smoking. Twenty-four percent

Table 1 Demographics.

Demographic ($n = 1448$)	Value
Median age at time of survey (years)	65
< 50	12.5%
50–59	20.8%
60–69	29.3%
≥ 70	35.5%
Male sex	54.3%
White race	90.2%
Cigarette use	
Current	3.2%
Former	65.7%
Never	30.7%
Gastroesophageal reflux disease	44.2%
Exposures	
Polluted air	30.2%
Asbestos	22.4%
Molds	19.7%
Pesticides, chemicals, fertilizers	10.5%
Dusts, fibers	10.8%
Family history of pulmonary fibrosis	11.9%
Year of diagnosis	
< 2000	23.9%
2000–2001	24.8%
2002–2003	39.9%
2004	10.2%

(23.9%) were diagnosed with pulmonary fibrosis prior to 2000.

A diagnosis of gastroesophageal reflux disease was reported by 44.2% of respondents. A wide variety of potential environmental exposures were reported by a minority. A family history of pulmonary fibrosis in at least one blood relative was reported by 11.9%. The mean age of diagnosis for relatives was 63.5 years.

Education and Resources

Sixty-four percent of respondents (63.5%) either somewhat or strongly agreed with the statement that there was a clear lack of information and resources about pulmonary fibrosis available at the time of their diagnosis (Table 2). Slightly over half (51.2%) reported being generally or very well informed about treatment options for pulmonary fibrosis at the present time. Fewer reported being generally or very well informed regarding the benefits of pulmonary rehabilitation (38.7%), managing supplemental oxygen (39.0%), nutrition (42.5%) and the risks and benefits of lung transplantation (32.5%). More recently diagnosed and older respondents reported being significantly less well informed about these issues (Table 2).

Experience with Diagnosis

Fifty-five percent (54.6%) of respondents reported at least a 1-year delay between the earliest indications of a potential breathing problem and the diagnosis of pulmonary fibrosis. Initial incorrect diagnosis was reported frequently: bronchitis (18.5%), asthma (14.1%) chronic obstructive

pulmonary disease (7.5%), emphysema (6.6%) heart disease (5.0%). Thirty-eight percent (38.2%) of respondents reported seeing 3 or more physicians before the diagnosis of pulmonary fibrosis was established. Fifty-three percent (53.2%) sought a second opinion. Eighty-four percent (84.4%) consulted a pulmonologist at some point during their diagnostic evaluation. Sixty-four percent of respondents (64.4%) reported being questioned about a family history of lung or respiratory disease. Only 27.5% recall having been told that genetic factors may predispose to pulmonary fibrosis.

Half of all respondents (50.2%) reported having a surgical lung biopsy. Surgical lung biopsy was less commonly reported the more recent the diagnosis. A minority reported having undergone bronchoscopy (21.5%).

Experience with Treatment

Overall, 74.7% of respondents reported current pharmacologic therapy for pulmonary fibrosis. The most common reasons given for not receiving pharmacologic therapy were fear of side effects (25.9%), ineffectiveness of therapy (22.7%), no treatment prescribed (23.3%), or early or stable disease (21.6%). The percentage of respondents reporting pharmacologic therapy was significantly less the more recent the diagnosis (Fig. 1).

Fifty percent of respondents (49.8%) reported currently taking prednisone for the treatment of pulmonary fibrosis. Another 20.9% had been prescribed prednisone in the past. A minority reported receiving cyclophosphamide (3.2% current, 5.8% former), azathioprine (10.2% current, 8.7% former) or colchicine (6.0% current, 6.5% former). Interferon gamma 1–b use was reported by 28.5% of respondents

Table 2 Patient education and resources.

Group (n)	Agree there was a lack of information/resources on pulmonary fibrosis at time of diagnosis (n)	Generally or very well-informed about (n)				
		Treatment	Rehab	Oxygen	Nutrition	Transplant
Total (1448)	63.5% (919)	51.2% (741)	38.7% (560)	39.0% (565)	42.5% (615)	32.5% (471)
<i>Year of diagnosis</i>						
<2000 (347)	68.3% (237)	55.0% (191)	47.0% (163)	48.1% (167)	48.4% (168)	40.3% (140)
2000–2001 (359)	63.0% (226)	58.2% (209) ^d	43.2% (155)	43.7% (157)	45.7% (164)	40.1% (144)
2002–2003 (578)	61.4% (355) ^a	49.1% (284)	34.4% (199) ^e	33.0% (191) ^f	38.6% (223) ^f	26.6% (154) ^e
2004 (147)	65.3% (96)	32.0% (47) ^e	24.5% (36) ^e	27.9% (41) ^f	34.0% (51) ^f	18.4% (27) ^e
<i>Age of time of survey</i>						
<50 (181)	75.1% (136) ^b	46.4% (84)	40.3% (73)	37.0% (67)	43.1% (78)	38.7% (70)
50–59 (301)	68.1% (205) ^c	53.5% (161)	38.2% (115)	35.9% (108)	46.8% (141)	37.2% (112)
60–69 (424)	61.6% (261)	54.0% (229)	40.6% (172)	41.7% (177)	45.0% (191)	35.8% (152)
≥70 (514)	58.8% (302)	49.6% (255)	37.0% (190)	40.1% (206)	38.1% (196) ^g	24.9% (128) ^h

a = $P < 0.05$ compared to year <2000.

b = $P < 0.05$ compared to ages 60–69 and ≥70.

c = $P < 0.05$ compared to age ≥70.

d = $P < 0.05$ compared to year 2002–2003.

e = $P < 0.05$ compared to all other years.

f = $P < 0.05$ compared to year <2000 and 2000–2001.

g = $P < 0.05$ compared to age 50–59 and 60–69.

h = $P < 0.05$ compared to all other ages.

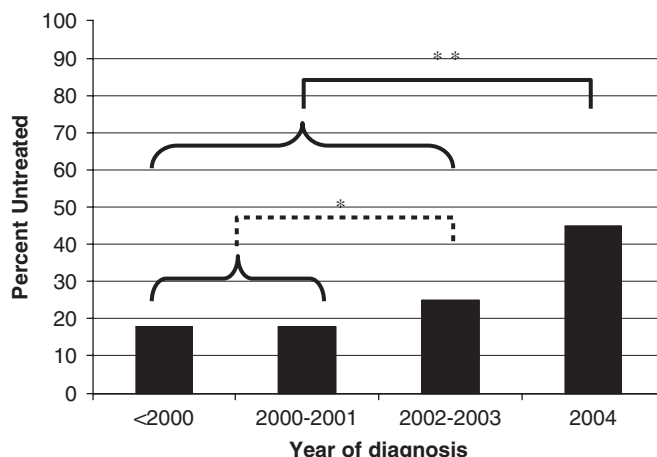


Figure 1 Percentage of respondents reporting no pharmacologic therapy by year of diagnosis. Significantly more respondents report no pharmacologic therapy for their disease the more recent the diagnosis (** $P < 0.01$, * $P < 0.05$).

(20.9% current, 7.7% former). A number of respondents (24.4%) reported the use of herbs or nutritional supplements.

Oxygen use was reported by 60.9% of respondents. Pulmonary rehabilitation and physical therapy referrals were reported by a minority of patients (31.8% and 23.9%). Behavioral health counseling referrals were uncommonly reported. Transplantation was discussed with the majority of respondents (58.7%).

Discussion

The results of this sample survey provide important insight into the experiences and attitudes of patients with pulmonary fibrosis. Despite continued attempts by the pulmonary community to clarify issues surrounding the diagnosis and treatment of this condition, including a joint ATS/ERS consensus statement on the diagnosis and management of IPF published in 2000,² this survey suggests there is a continued failure on the part of clinicians to educate patients to their diagnosis. There are several reasons why patients may feel so ill-informed. Despite the best effort of the medical community to educate patients, many patients will require time to accept the diagnosis and repetition of information regarding its diagnosis and management to fully understand the implications. The perception of being uninformed may result more from these issues than a failure on the part of the treating physician. Nonetheless, the fact that over half of survey respondents reported a clear lack of information about pulmonary fibrosis at the time of diagnosis and felt poorly informed about important non-pharmacological management options is unacceptable.

Two other findings deserve mention. First, approximately half of respondents reported undergoing a surgical lung biopsy for the diagnosis of their condition. While these data are patient-reported and therefore open to criticism, this percentage is in keeping with data that suggest approximately half of patients with IPF can be diagnosed clinically.^{8,9} Second, the majority of respondents (71%) reported current or prior treatment with prednisone. Only a minority reported receiving cytotoxic agents such as cyclophospha-

mid or azathioprine. Approximately 30% reported current or prior treatment with interferon gamma 1-b. Although these results may be due to treatment bias in our responding population, they suggest that the ATS/ERS recommendation for combined therapy with prednisone and cytotoxic agents may not be commonly followed, and that many patients are receiving off-label investigation agents. Third, the percentage of respondents reporting no pharmacologic therapy increased significantly the more recent the diagnosis. Patients and physicians appear to be deciding to forego medical therapy more frequently than in the past. The data do not offer an explanation for this observation.

This project represents the first large-scale survey of patient experiences and perceptions regarding the diagnosis and treatment of pulmonary fibrosis published to date. Two surveys of physicians have been published in the last 20 years. A small survey of physicians was conducted in 1989 to describe approaches to the management of interstitial lung disease, but only included 25 respondents.¹³ More recently, a survey of physicians in the United Kingdom compiled 588 patients with IPF (termed cryptogenic fibrosing alveolitis) and described their presentation, diagnosis and initial management.¹⁴ Importantly, the case definition of cryptogenic fibrosing alveolitis required only chest radiographic findings but not computed tomography or pathology, thereby likely including other histopathologic expressions of idiopathic interstitial pneumonia. The authors reported that the vast majority (88%) of patients were diagnosed without surgical lung biopsy, and nearly half (48%) of patients were never treated.

There are important limitations to this current survey that limit and qualify any conclusions drawn. First and foremost are issues of sampling. CPF members are self-identified as having pulmonary fibrosis; no confirmation of the diagnosis can be obtained. It is important that the results of this survey not be directly extrapolated to the population of patients with well-defined IPF. In addition, this survey employs non-probability sampling, which introduces potential sampling bias. Unfortunately, probability sampling of pulmonary fibrosis patients is impossible as there is no central registry from which to sample. Both of these issues impact the generalizability (i.e. external validity) of the results.

Second, a large number of non-responses can bias the results of a sample survey because the population of non-responders may have substantial differences from the population of responders (e.g. younger, less dissatisfied, less educated). In this sample survey, the response rate approached 50%. While this percentage is higher than many medical opinion surveys report, a substantial number of non-respondents remain.

Third, the information obtained from respondents may be inaccurate (misinformation bias). Respondents may not correctly understand the subject matter of the question (e.g. transbronchial lung biopsy may be confused for a surgical lung biopsy), or respondents may not recall events accurately. Finally, this survey does not formally assess patients' quality of life and cannot comment on the impact of patient perceptions on this important outcome measure.^{15,16}

These potential sources of error are common to surveys and cannot be completely eliminated. Nonetheless, this survey's results suggest important findings, most prominently, an unacceptable failure in patient education regarding the diagnosis of pulmonary fibrosis. These results should prompt academic and practicing physicians, patients, and patient advocates to work together to better define and disseminate information about pulmonary fibrosis, and motivate providers to redouble efforts to educate their patients with pulmonary fibrosis about the diagnosis and management of their disease.

Appendix A. Supplementary Materials

Supplementary data associated with this article can be found in the online version at [doi:10.1016/j.rmed.2006.10.002](https://doi.org/10.1016/j.rmed.2006.10.002).

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