ORIGINAL RESEARCH LETTER

Healthcare pathway and patients' expectations in pulmonary fibrosis

To the Editor:

In a survey of European idiopathic pulmonary fibrosis (IPF) patient advocacy groups, Bonella *et al.* [1] highlighted inequalities in IPF care across Europe. The European IPF patient charter summarises patients' priorities and underscores the frequency of misdiagnosis, of delayed referral to excellence centres, and the need for education of general physicians and pulmonologists about IPF.

To better understand the healthcare pathway and expectations of patients with pulmonary fibrosis, a study was conducted in 24 randomly selected patients with pulmonary fibrosis identified from the cohorts of academic centres in four French regions (Rhône-Alpes, Languedoc-Roussillon, Ile-de-France and Midi-Pyrénées). Following oral consent, semi-structured interviews of about 1 h each, with pre-defined questions for each topic of interest, were conducted by a single professional nurse with specific training as an interviewer as well as knowledge in IPF. Interviews were conducted at the home of each patient, independent of the medical team, and blinded to medical data. The topics covered by the interviews systematically included the healthcare pathway and patient journey, the feelings and experience of living with the disease, and expectations regarding improvement in patient care. The interviews were audio-recorded, transcribed verbatim, and the data were analysed through framework analysis. The study was evaluated by an ethics committee of research, which confirmed that it did not raise ethical concerns. As no additional investigation was performed for this study and patients who did not consent declined the interview, written consent was waived for interviews.

The patients (14 male and 10 female), with a mean age of 68.0±10.4 years, had been diagnosed with IPF (n=15), idiopathic nonspecific interstitial pneumonia (n=1), or pulmonary fibrosis associated with a specific context (connective tissue disease in 5, fibrotic hypersensitivity pneumonitis in 3).

All patients reported difficulties in accessing a centre with expertise in interstitial lung disease (ILD) and a delay in obtaining a secure diagnosis (more than 1 year in 14/24 cases), with a need to see several pulmonologists before being referred to the excellence centre (16/24 patients), in most cases due to local physicians having little knowledge of to whom and how to refer their patients. Although the specific delays were not assessed, the delay in accessing the excellence centre was felt by patients to be due equally to waiting until seeking medical advice and to insufficient coordination between the physicians involved (general physician, local pulmonologist, and hospital specialist).

Importantly, 21/24 patients (87%) expressed intense relief when eventually seen at an excellence centre, based on there being adequate information about their rare lung condition; confidence that a secure diagnosis was made; up-to-date knowledge in the availability, indication and management of treatments; and satisfaction that disease follow-up was organised by the centre together with the local pulmonologist. However, 18/24 patients wished their local physicians were more actively integrated into patient care, and 16 patients reported difficulties in the circulation of relevant information between the general physician/local pulmonologist and the excellence centre, despite medical reports being available.

Patients' expectations from the excellence centres included expert medical management, with a secure diagnosis, individualised therapy, and a reassuring organisation of follow-up, with adequate coordination between the ILD centre and the local physicians. Earlier referral to excellence centres was considered crucial to the patients' experience. Patients expressed the need for an improved process of announcement of the diagnosis, with sufficient time dedicated, adequate material settings, a sensitive psychological

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approach and comprehensive information covering the disease and its anticipated evolution. They also asked to be informed about how they could be accompanied in the healthcare system throughout the disease course. Being seen at the excellence centre contributed to their feeling of being supported and to an improvement in the quality of life, even at an advanced stage of disease. Several patients mentioned that only at the excellence centre was the issue of end of life addressed and their questions answered. Overall, the patients expressed a need for global support, including social and psychological support. No difference was seen between patients with IPF and those with other kinds of pulmonary fibrosis.

Patients expressed interest in being informed about drug therapy and side effects, but they generally did not wish to take an active part in the treatment decision. In addition, 18/24 patients needed stakeholders other than the excellence centre to better support them in the following priority domains: information about the disease and how to cope on a daily basis; better access to non-drug therapy, from physiotherapy and education to the use of ambulatory oxygen supplementation; psychological support and the opportunity to speak to persons outside the medical team; social and financial support and guidance in the administrative procedures needed to seek support; and supportive care, management of symptoms and end-of-life support.

This survey clearly highlights that patients' expectations lie far beyond just medical management of the disease and drug therapy, with a requirement for more comprehensive management and global support. The results further underscore that the patient journey remains a long and complex path in most cases, consistent with what is known in rare and "orphan" diseases [2]. Although much attention is currently directed towards IPF, patients with other progressive pulmonary fibrotic diseases have similar needs. This survey highlights an important unmet need in patients, especially given that it was observed in France, where successive national plans for rare diseases have generally organised the care of these diseases well and identified specialised centres, including those for rare pulmonary diseases (www.maladies-pulmonaires-rares.fr). Such unmet needs may be even higher in countries where the care of rare diseases is less organised. Patients' testimonies [3] and other studies in IPF [4–7] and other rare pulmonary diseases [8–10] have produced somewhat comparable results, despite those interviews being more focused on patients' experience and coping [4] rather than on the patient journey, but access to centres of excellence was identified as an unmet need. In a survey of 55 patients with IPF from five European countries, SCHOENHEIT *et al.* [5] showed that patient satisfaction with medical care and disease education was higher among patients who were receiving care at a recognised centre of excellence [5].

Interestingly, patients expressed only modest interest in discussing the choice of drug and the indication of treatment, as they generally felt that they did not have sufficient knowledge regarding such specific questions to take an active part in the treatment decision. This supports the notion that patients and patient associations should be involved upstream at the stage of guideline writing [11] in addition to that of the individual patient–physician interaction and treatment decision.

Patients adapted to the illness according to their personality and disease stage. Some could be considered as "actors" of their own management, whereas others were "protected" from problems and decisions by their relatives. A third category of patients had a psychological profile of "victim", with little personal autonomy and socio-affective support. Such patients' "typologies", identified by the survey, may be an interesting starting point for better tailored patient support. For example, "actors" request tools to help organise their management, "protected" may benefit from advice given to their relatives and caregivers, and "victims" would need external psychosocial support. Whether these profiles may help in adapting support to patients' needs requires further evaluation.

This study has limitations. Only questions addressed in semi-structured interviews were discussed. For example, the possible input of patients in trial design [12] was not addressed. All patients in this study were eventually diagnosed with pulmonary fibrosis in a specialised academic centre, which might have introduced a selection bias in favour of patients with the longest and most complex patient journey. Healthcare pathways were not analysed according to the geographic origin of the patients. We consider, however, that the patients' experiences were representative of routine management.

In conclusion, the results of this study underscore the need for an earlier referral to excellence centres and for an improved coordinated network of care for patients with progressive pulmonary fibrosis that can provide comprehensive support and disease management.

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