Patients Suffering from a Chronic, Irreversible Illness: A Novel Study on the Psychological Intervention out of the Hospital

David Rudilla, Amparo Oliver, Laura Galiana,
Claudia Valenzuela, and Julio Ancochea

Introduction

Since the 1980s, health psychology has been interested in identifying psychological aspects of patients suffering from rare, chronic, and advanced-stage diseases. This would be the case with chronic obstructive pulmonary disease (COPD), diffuse interstitial lung disease (DILD), and idiopathic pulmonary fibrosis (IPF). When compared with cancer patients, those suffering with IPF expressed similar or more severe symptoms and care needs [1]. However, the interventions in these patients were minor and limited to few variables, ignoring many other key issues in the perception of quality of life associated with the disease, and are presented in the Results section of this chapter.

The prevalence and incidence of idiopathic pulmonary fibrosis (IPF) varies in its definition and geographic location, with a prevalence of 1.25–63 cases and an annual incidence of 0.22–17.4 cases per every 100,000 inhabitants, with more cases in North America than in Europe [2]. It is the most frequent entity within the group of the National Institute of Immunology (NII), with an estimated prevalence in Spain of 13 cases per 100,000 women and 20 cases per 100,000 men, therefore affecting approximately 7000–12,000 patients, with a growing tendency [3, 4].

IPF is a chronic, irreversible, fibrosing lung disease with an unfavorable prognosis. The clinical course of patients with IPF varies from asymptomatic patients to those who have a progressive loss of lung function and on to others who may experience very rapid deterioration. Some patients may present episodes of sudden worsening, with acute respiratory failure, which we call acute exacerbations [5]. The combination of the poor prognosis, the uncertainty of the course of the disease, and the severe burden of the symptoms has a great impact on the quality of life both for patients and their relatives [6–8]. Although in recent years, with the appearance of two antifibrotic treatments, studies have been conducted, most of these trials focus on the modification of the disease measured by physiological parameters such as lung function. However, in many patients with IPF, there is no clear correlation between physiological parameters and outcome measures reported by patients regarding how they feel or cope with daily activities.

D. Rudilla
Pneumology, La Princesa University Hospital,
Madrid, Spain

Air Liquide Healthcare, Madrid, Spain
e-mail: david.rudilla@airliquide.com

A. Oliver (✉) · L. Galiana
University of Valencia, Valencia, Spain
e-mail: amparo.oliver@uv.es; laura.galiana@uv.es

C. Valenzuela · J. Ancochea
Pneumology, La Princesa University Hospital,
Madrid, Spain

Autonomous University of Madrid, Madrid, Spain
e-mail: j.ancochea@separ.es

M. Silbermann (ed.), *Palliative Care for Chronic Cancer Patients in the Community*,
https://doi.org/10.1007/978-3-030-54526-0_11

Table 1 Results for the basal line ($n = 56$)

	Min.	Max.	Mean	SD
Age	40	81	63.55	9.55
Anxiety	3	19	11.17	4.31
Depression	2	17	8.42	4.17
Demoralization	2	20	11.33	4.69
Quality of life	2	7	4.14	1.31
Spirituality intrapersonal	4	12	9.19	1.99
Spirituality interpersonal	2	12	10.42	2.03
Spirituality transpersonal	1	8	5.71	1.83
Threaten dignity	4	27	14.39	6.29
Preserved dignity	23	45	35.62	5.09
Dignity	35	72	50.01	9.31
Resilience	4	20	10.78	4.56
Confidential support	13	30	24.19	5.35
Affective support	12	25	19.82	3.84
Social support	25	55	44.01	8.95

More recent studies have examined the psychological needs of IPF patients and their families. Table 1 shows the studies that have explored the psychological aspects of the patient with IPF. Simultaneously, at the international level, IPF is becoming the leading indication for lung transplantation [9], and this could also introduce psychological symptoms associated with the uncertainty of this process. Anxiety and depression are known to frequently occur in patients with chronic respiratory diseases [8, 9]. In COPD, the intensity of dyspnea reported by patients is directly related to depression and the impairment of functional status [10].

Patients with diffuse interstitial lung disease (DILD) also report inadequate information regarding the disease process, an imprecise diagnosis, unsatisfactory treatment, and/or unacceptable side effects associated with therapy and poorly controlled symptoms of progressive illnesses. Ryerson et al. [6], in a study of patients suffering from DILD using the Center for Epidemiologic Studies Depression Scale (CESD), found that even a moderate degree of dyspnea experienced by patients with DILD resulted in a high incidence of depression. Therefore, depression and anxiety require special attention when assessing chronic lung diseases.

Pulmonologists have frequently emphasized their clinical impressions of the role that psychological components play in different respiratory pathologies. Holland et al. [11] corroborated the findings of Ryerson and his collaborators regarding the prevalence of anxiety and depression in DILD. They observed a growing presence of anxiety and depression across the spectrum of this disease without predilection for any particular sub-type.

Participating in a pulmonary rehabilitation program improves both mood and functional capacity, and enrolling in a local IPF support group may help reduce social isolation [12]. A palliative care program for patients and their caregivers focused in the home environment demonstrated significant improvements in health-related quality of life and respiratory symptoms [13]. However, studies that center on the relationship between psychological variables and the clinical manifestations of pulmonary diseases are still scarce [14–17]. Literature has established the presence of clinical symptoms of anxiety and depression in 21–50% of patients diagnosed with diffuse interstitial diseases, almost all with IPF. Recently, two papers have addressed the clinical impact of IPF on anxiety and depression. The first was exploratory in IPF patients and their relatives and found that a high incidence of anxiety was reported, and most considered psychological support essential [16].

The second study showed that the prevalence of anxiety and depression in patients with IPF was 25.9% and 21.4%, respectively [17]. However, most studies that address psychological variables are inconclusive, basically due to problems with sample size [18, 19], or because they focus solely on assessing the physical benefits of respiratory rehabilitation programs [20].

In the reviewed studies, patients with IPF were involved in almost all domains of life, but the areas most impacted were related to physical functioning, symptoms, and daily autonomy. The results significantly correlated with indicators used to assess the severity of IPF, with both physiological measurements and subjective indicators such as dyspnea. The degree of dyspnea was also associated with a depressive mood [21–24]. Duck et al. [25] identified three main themes in their interviews with patients with IPF: “fighting for a diagnosis,” “loss of the life that they had previously led,” and “living with idiopathic pulmonary fibrosis." Patients reported that they struggle to cope with a disease that limits their lives and progresses rapidly without sufficient treatment or support structures. These findings are in line with those reported by Sampson [26], in which patients diagnosed with IPF had a clear understanding of their prognosis, but little understanding of how their disease would progress or be managed.

There are few investigations of palliative care in advanced stages of IPF [18, 27–31], although problems related to symptom control and the deterioration of patients' quality of life are similar to those that occur, for example, in palliative cancer patients [32]. Thus, we have no information on the psychological profile of patients with IPF at the end of life: the presence of demoralization syndrome, spiritual needs, or perception of dignity. These three constructs, together with emotional distress, are most characteristic of patients at the end of life, having been the focus of oncological research but with less attention given to the importance of non-oncology issues.

The term "demoralization" is an alternative to the diagnosis of depression that seems to be more prevalent in patients with advanced chronic disease where discouragement, loss of meaning of life, helplessness, hopelessness, and feelings of failure characterize the mood of the patient [33]. Unlike the depressed patient, the patient with demoralization has no anhedonia and is able to enjoy some things. Mirjam and his collaborators [34] have recently described the ABCDE plan, which involves a comprehensive approach to the patient and which considers psychological intervention to be at the same level as the treatment of physical symptoms. This approach considers the participation of different professional profiles (pulmonologists, nurses, psychologists, etc.) a necessity.

Taking into account all these antecedents and needs of patients with IPF, this study presents the results obtained after a structured ambulatory psychological intervention.

Participants and Methods

Procedure and Design

After obtaining the permits from La Princesa University Hospital's Ethical Committee and patients' informed consents, patients diagnosed with IPF began psychological treatment at the Pulmonology Unit. For ethical reasons, as stipulated by the committee, a pre-test/post-test, non-randomized trial was carried out and no control group was set up, but everyone was offered to participate.

Pre-evaluations were carried out at the beginning of Session 1, and post-intervention evaluations were carried out at the end of the last session. The interventions took place between November 5, 2017, and June 7, 2018, in the consulting room of the psychology department of the DILD Unit, located in the Pulmonology Unit of the La Princesa University Hospital. The intervention consisted of three sessions of brief psychotherapy and focused on the essential dimensions of the person (dignity, demoralization, hopelessness, and spirituality), using counseling as a basic therapeutic methodology. The contents of the sessions were:

- 1st session: Who am I?
- 2nd session: Hope and demoralization
- 3rd session: Spirituality and dignity

Interventions were held every 2 weeks. A flowchart describing the study can be found in Fig. 1.

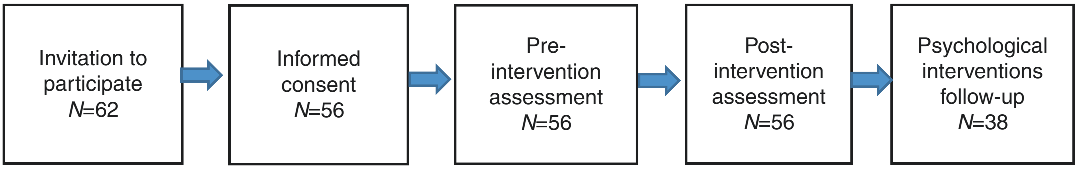

Fig. 1 Interventions

Sampling

The participants were patients diagnosed with IPF and were being followed-up by the Pulmonology Unit of La Princesa University Hospital.

The inclusion criteria were:

(a) Patients diagnosed with IPF being followed-up by the unit
(b) Adult patients (over 18 years old)
(c) Informed consent
(d) Survival expectancy greater than 6 months

The exclusion criteria were:

(a) Existence of a pact of silence (not knowing diagnosis or prognosis)
(b) Cognitive impairment (comprehension/expression problems)

Instruments

In addition to registering socio-demographic and clinical variables, the following instruments were administered:

(a) Hospital Anxiety and Depression Scale (HADS) [35, 36]. This instrument assesses affective disorders in the non-psychiatric hospital setting. It consists of 14 items with a four-point Likert response format (ranging from 0 to 3): seven for each subscale, anxiety and depression. The score for each subscale is 21 points, which is achieved with the sum of the items corresponding to each symptom. The reliability of the current application is 0.75 for anxiety and 0.71 for depression.
(b) Brief Demoralization Scale [37] that evaluates the demoralization syndrome by means of five brief elements with a Likert response that varies from none to a lot. Reliability measured by application is 0.85.
(c) Dignity Assessment Questionnaire (PPDS) [38]. Brief instrument to measure dignity in patients with and without palliative cancer, with an alpha in present application of 0.72. It consists of eight elements with a response scale of 0 (none) to 9 (a lot). It consists of two factors: preservation of dignity and threat of dignity.
(d) Spirituality (GES) [39]. Spirituality is associated with quality and meaning in life; it is a complex construct with more than 13 components [40] and is clearly different from religion or faith. The measurement tool consists of eight items that are answered with a five-point Likert scale ranging from 0 (nothing) to 4 (a lot). It includes three factors: intrapersonal, interpersonal, and transpersonal dimensions. The original Cronbach's alpha for this scale is 0.86
(e) Brief Resilience Scale [41]. This four-item scale was adapted and validated in Spain [42] with an internal consistency of 0.79 and in its current application has reached 0.92. Each item is scored on a five-point scale from 1 (strongly agree) to 5 (strongly disagree), and the highest scores reflect a greater resilience.
(f) Duke-UNC [43] Functional Social Support Questionnaire. Quantitative evaluation questionnaire of perceived social support relating to two aspects: people with whom intimate feelings can be communicated (confidential support) and people who express positive feelings of empathy (emotional or affective support). It collects values that refer to both confidential support and affective support. Each aspect is evaluated with a Likert scale with five options from 1 (much less than I want) to 5 (as much as I want). Overall reliability is 0.80.

Quality of life. Two elements of the EORTC-QLQ-C30 [38, 44] were used, which constitute the factor of the Global Health Scale. QLQ-C30 is currently used in several types of studies of people with cancer and other advanced chronic diseases. The use of the complete questionnaire was rejected and, as physical deterioration is inevitable in patients undergoing symptomatic treatment, it is recommended to use only the aforementioned indicators in studies. These studies also included patients with advanced respiratory diseases, and no comparative differences from cancer patients were found. Reliability of this measure in our patients was 0.75

Analysis

The equivalence of groups was tested according to whether the patients were following treatment with an antifibrotic. For the quantitative variables, *t* tests were performed for comparison of means of independent groups, with the appropriate correction to the diagnosis of compliance with the assumption of homogeneity of variances. Regarding categorical variables, χ^2 tests were used.

In order to view the clinical map of the essential dimensions, bivariate correlations were calculated. To know the differences between the results in variables of each of the types of intervention, *t* tests of repeated measures were carried out. In addition, the same tests were used to compare differences between interventions, except for independent samples. In all tests, the size of the associated effect was calculated. In the case of the effect size for the *t* tests, Cohen's d was calculated to determine the standardized difference between every two means under comparison. Cohen's d has values from 0 to infinity, with interpretation of small values at around 0.20, medium size around 0.50, and large size from 0.80 [45].

Results

The final sample consisted of 56 patients. 42% were women and 58% were men. 46% of the patients had been diagnosed more than 12 months ago, and 60% were on antifibrotic treatment. The rest of the characteristics can be consulted in Table 1.

When dividing the sample into two groups, depending on whether they were being treated with an antifibrotic, no significant baseline differences were found between both groups in gender ($\chi^2(2) = 1.373$; $p = 0.503$), marital status ($\chi^2(6) = 4.543$; $p = 0.604$), level of studies ($\chi^2(6) = 11.205$; $p = 0.082$), employment situation ($\chi^2(8) = 3.126$; $p = 0.926$), or diagnosis time ($\chi^2(8) = 14,494$; $p = 0.070$). Nor did Student's *t* test with independent samples correcting the error inflation find difference in age ($t(53) = 1.147$; $p = 0.256$).

Following, in Table 2, we appreciate the amount of significant correlations between key variables involved in the process of caring with dignity and also the intensity of these associations.

Regarding the changes after the interventions (Table 3), the *t* tests for paired samples showed statistically significant differences in all the variables ($p < 0.004$) except in depression ($p = 0.05$), transpersonal spirituality ($p = 0.075$), and confidential social support ($p = 0.465$). Bonferroni adjustment was applied to correct error inflation due to the simultaneous pairwise comparisons made.

Following these lines, Fig. 2 below more intuitively shows, in a visual display, the changes observed in the target variables after the intervention.

As shown in Fig. 2, as expected a priori, in some of the comparisons after the psychological intervention, the results are better except in depression, where there was an increase, although this is not significant. They also increase spirituality, quality of life, dignity, resilience, and social support. Regarding effect sizes, they were of great magnitude ($d > 0.8$) in quality of life and preserved dignity and of medium size ($0.8 > d > 0.5$) in intrapersonal spirituality and affective support.

Finally, several analyses of variance were carried out to test for the effect of using an antifibrotic. The results indicate that the changes already found are independent of the type of treatment being followed, with or without an antifibrotic (see Table 4).

Table 2 Relationships among the whole set of variables in basal line

	1	2	3	4	5	6	7	8	9	10	11	12	13
1. Anxiety	1												
2. Depression	0.629[a]	1											
	0.000												
3. Quality of life	−0.678[a]	−0.552[a]	1										
	0.000	0.000											
4. Demoralization	0.521[a]	0.469[a]	−0.332[b]	1									
	0.000	0.000	0.012										
5. Spirituality Intra	−0.270[b]	−0.189	0.284[b]	−0.223	1								
	0.044	0.163	0.034	0.099									
6. Spirituality Inter	−0.274[b]	−0.022	0.399[a]	−0.297[b]	0.561[a]	1							
	0.041	0.872	0.002	0.026	0.000								
7. Spirituality Trans	−0.338[b]	−0.425[a]	0.417[a]	−0.467[a]	0.343[a]	0.491[a]	1						
	0.011	0.001	0.001	0.000	0.010	0.000							
8. Preserved Dig.	−0.409[a]	−0.312[b]	0.454[a]	−0.329[b]	0.389[a]	0.321[b]	0.286[b]	1					
	0.002	0.019	0.000	0.013	0.003	0.016	0.033						
9. Resilience	−0.074	0.100	0.202	−0.114	0.196	0.339[b]	−0.103	−0.200	1				
	0.590	0.463	0.135	0.401	0.147	0.011	0.451	0.140					
10. Threatened Dig.	−0.387[a]	−0.467[a]	0.443[a]	−0.449[a]	0.160	−0.008	0.153	0.331[b]	0.131	1			
	0.003	0.000	0.001	0.001	0.238	0.955	0.260	0.013	0.335				
11. Dignity	−0.485[a]	−0.486[a]	0.547[a]	−0.483[a]	0.321[b]	0.170	0.260	0.770[a]	−0.020	0.857[a]	1		
	0.000	0.000	0.000	0.000	0.016	0.209	0.053	0.000	0.881	0.000			
12. Conf. Support	−0.386[a]	−0.062	0.295[b]	−0.089	0.112	0.479[a]	0.398[a]	0.144	0.235	−0.096	0.014	1	
	0.003	0.648	0.028	0.512	0.411	0.000	0.002	0.289	0.081	0.483	0.918		
13. Affective Sup.	−0.325[b]	−0.004	0.252	0.068	0.185	0.461[a]	0.371[a]	0.209	0.065	−0.181	−0.008	0.892[a]	1
	0.015	0.975	0.061	0.619	0.172	0.000	0.005	0.121	0.634	0.182	0.953	0.000	
14. Social Support	−0.370[a]	−0.039	0.284[b]	−0.024	0.146	0.485[a]	0.397[a]	0.176	0.169	−0.135	0.005	0.981[a]	0.963[a]
	0.005	0.775	0.034	0.859	0.282	0.000	0.002	0.194	0.214	0.322	0.971	0.000	0.000

Abbreviations: *Dig*. Dignity, *Conf*. Confidential, *Sup*. Support

[a] $p < 0.01$

[b] $p < 0.05$

Table 3 Results along the intervention

	Pre		Post					
	M	SD	M	SD	*t*	df	*p*	d
Anxiety	11.17	4.31	9.05	2.51	3.06	55	0.003	0.40
Depression	8.42	4.17	9.82	2.62	−2.00	55	0.050	0.27
Demoralization	11.33	4.69	9.82	2.62	−3.10	55	0.003	0.41
Quality of life	4.14	1.31	7.00	2.45	7.16	55	0.000	0.96
Spirituality intra	9.19	1.99	4.81	0.99	−5.33	53	0.000	0.72
Spirituality inter	10.42	2.03	11.27	1.91	−3.64	53	0.001	0.49
Spirituality trans	5.71	1.83	11.42	0.83	−1.81	53	0.075	0.25
Threaten dignity	14.39	6.29	11.33	4.09	3.28	55	0.002	0.44
Preserved dignity	35.62	5.09	42.41	1.86	−10.75	55	0.000	1.44
Dignity	50.01	9.31	53.75	5.40	−3.04	55	0.004	0.41
Resilience	10.78	4.56	12.92	4.27	−1.15	53	0.255	0.16
Social support	44.01	8.95	46.54	3.05	−2.04	49	0.046	0.29
Conf. support	24.19	5.35	23.62	1.39	0.73	55	0.465	0.10
Affective support	19.82	3.84	22.92	2.01	−5.19	49	0.000	0.73

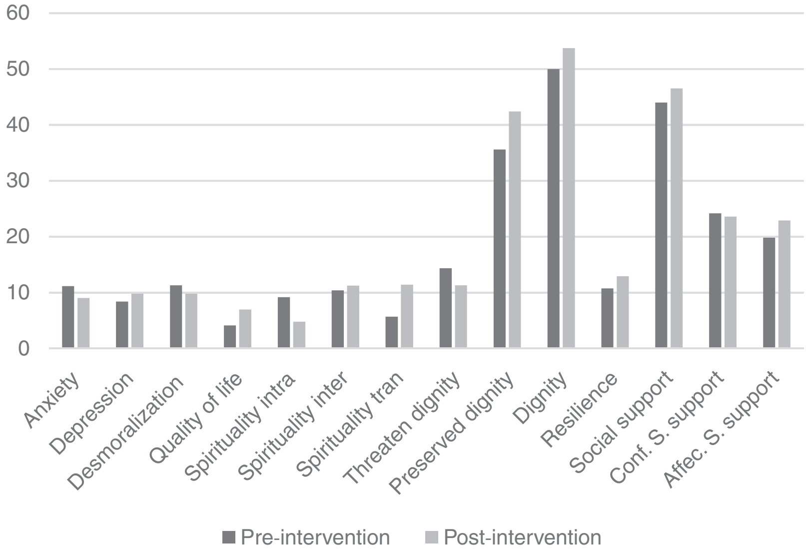

Fig. 2 Observed change in variables after the intervention

Table 4 Analysis of variance testing differences by type of treatment

Outcome	df	*F*	*p*	η^2
Anxiety	1	1.320	0.255	0.024
Depression	1	0.649	0.424	0.012
Demoralization	1	0.404	0.528	0.007
Quality of life	1	3.540	0.065	0.062
Spirituality intra	1	0.861	0.358	0.016
Spirituality inter	1	0.356	0.553	0.007
Spirituality trans	1	3.168	0.081	0.057
Dignity	1	0.179	0.674	0.003
Preserved dignity	1	0.036	0.851	0.001
Threatened dignity	1	0.175	0.678	0.003
Resilience	1	1.619	0.209	0.029
Confidential social support	1	0.668	0.417	0.012
Affective social support	1	0.275	0.602	0.006
Social support	1	0.000	0.995	0.000

Discussion

The objective was to evaluate the effectiveness of a structured psychological intervention to improve the quality of life of patients diagnosed with IPF. In general, the results showed that a brief psychotherapy intervention with content focused on the essential dimensions of the individual was beneficial for patients with IPF. As it happens in interventions of this style, anxiety levels were reduced in patients with early palliative profile [46–49]. The results of this study, however, do not show such improvement in terms of depression. The effects found in this work have not been statistically significant and with small sizes. These results are not far from those consulted in the depression literature that have been found when intervening in palliative patients [47, 48]. Possibly due to the same explanation: the depressive mood is explained by a demoralization and not by a depressive episode.

Along these lines, Houmann, Rydahl-Hansen, Chochinov, Kristjanson, and Groenvold [50] and Rudilla et al. [48] also found evidence that depression levels did not improve after interventions that address existential distress such as Dignity Therapy, but that they were even higher. These types of interventions in palliative patients do not seem to improve mood, although anxiety symptoms do.

Regarding the quality of life, psychotherapy showed positive and large effects on this variable. This result matches with what is proposed by Juliao et al. [47], Rudilla [48], and Chochinov et al. [51], among others. Finally, regarding the emotional distress caused by the lack of dignity, or the improvement of the spiritual dimension, results of this study also point to an improvement after the intervention, although only for some of the dimensions of spirituality (intrapersonal and interpersonal) and dignity and its dimensions. These results coincide with those found in different studies that use therapies of the essential dimension such as Dignity Therapy [47, 48, 52–54].

It is important to note that the results found that these benefits occurred both in patients treated with an antifibrotic and symptomatic treatment. There is a need for emotional support in patients with IPF.

In conclusion, it could be said that a brief psychotherapy session structured around the contents of existential anxiety has improved patients' quality of life, their dignity and spirituality, and also, to some extent, their emotional well-being. However, the fact that no previous studies aimed at patients with IPF have been carried out does not allow us to go in-depth in the elements for that success as there is no previous specific literature to compare it to. We can only provide some limited first or pilot evidence on the impact of this therapy in the medium term. It is still necessary to carry out follow-up measures to know if these changes are consolidated, as they suppose a personal resource in view of the progressive deterioration to which the patients with IPF are subjected, if maintenance is necessary, and, if so, what type. Nor have autonomy measures been taken that would allow us to know how psychotherapy impacts dependence.

For these reasons, it would be necessary for future research to address this problem with a larger sample size and clinical measures to study the possible replicability of the results found in this work. It is important to determine which interventions cover the needs of patients with IPF that have been neglected so far and, therefore, should be implemented by professionals. Another important aspect focuses on the quality of life of the palliative patient.

The evaluation carried out in this study of quality of life has been carried out with the global health status factor of the EORTC-QLQ-30, of simple application (thus avoiding the fierceness of evaluating with long and tedious questionnaires) and which has shown to be sensitive to studies with non-cancer palliative patients [48]. To include other questionnaires that determine quality of life through measuring symptoms in patients whose clinical deterioration implies a continuous loss supposes obtaining a negative result that may not reflect the true state of quality of life of the patient.

The evaluation of quality of life that focuses solely on the physical aspects neglects the possibility that patients can perceive a better quality of life in issues of a psycho-emotional nature and, therefore, can improve relationships with their relatives and others close to them [55]. The referred loss can result in worsening of mood (greater depression). It is important to emphasize these aspects when talking about "quality of life" in the patient with IPF.

Patients with serious illnesses tend to have great existential suffering, and in response to this need, specific interventions essentially focused on end of life (counseling, Kibo Therapeutic Interview [56]) have been developed. These types of interventions, in the context of the Spanish healthcare system, are not mandatory despite the recent commitment for increasing humanization of care. There are pathologies as adverse as lung cancer, such as IPF, in which protocoled treatments aimed at reducing existential suffering and emotional distress are being developed. These are rare or minor diseases; it is assumed from the start that they will conclude with a poor prognosis.

Advancements in the care of patients with serious illnesses must consider the entire person. The improvement of a treatment that increases survival rate implies that the set of treatments that affect the person, such as psychological and spiritual, be modified. In addition, each time the treatments are transferred to the patient's home environment, it is considered to be the best therapeutic space for the patient and his/her family. For this reason, the approach to the patient must have interventions that do not have the sole objective of admission or clinical worsening but should strengthen and provide coping strategies that allow the patient and his/her family to live day to day. It is necessary that health professionals' articulate mechanisms ensure that everyone (doctors, nurses, psychologists, patients, and caregivers) remembers that, before being patients, they are human beings.

By evidencing the usefulness of these interventions, we are attempting to stimulate a debate about a redesigning of end-of-life care protocols. Just as no one doubts the effectiveness of the case management nurse, approaches such as the intervention of the three psychotherapeutic sessions presented here are part of generic protocols for action. This has additional benefits such as avoiding treating groups of patients separately depending on their disease. By separating, it is inevitably labeled and sometimes even stigmatized, and with interventions like this, we are moving toward a more compassionate system of care, healthcare models that can be transformed into systems that honor the dignity of people, as was defended at the *International Conference on Improving the Spiritual Dimension of Whole Person Care: The Transformational Role of Compassion, Love, and Forgiveness in Health Care* in 2013 [57].

References

1. Callahan D. Death and the research imperative. N Engl J Med. 2000;324:654–6.
2. Nalysnyk L, Cid-Ruzafa J, Rotella P, Esser D. Incidence and prevalence of idiopathic pulmonary fibrosis: review of the literature. Eur Respir Rev. 2012;21:355–61.
3. Ancochea J, Xaubet A, Agüero R, editors. Fibrosis pulmonar idiopática. Madrid: Editorial Respira; 2015. p. 41–57.
4. Xaubet A, Ancochea J, Bollo E, Fernández-Fabrellas E, Franquet T, Molina-Molina M, et al. Normativa sobre el diagnóstico y tratamiento de la fibrosis pulmonar idiopática. Normativa SEPAR Arch Bronconeumol. 2013;49(8):343–53. https://doi.org/10.1016/j.arbres.2013.03.011.
5. Yount SE, Beaumont JL, Chen SY, Kaiser K, Wortman K, Van Brunt DL, et al. Health-related quality of life in patients with idiopathic pulmonary fibrosis. Lung. 2016;194(2):227–34. https://doi.org/10.1007/s00408-016-9850-y.
6. Ryerson CJ, Berkley J, Carrieri-Kohlman L, Pantilat SZ, Landefeld CS, Collard HR. Depression and functional status are strongly associated with dyspnoea in interstitial lung disease. Chest. 2011;139(3):609–16. https://doi.org/10.1378/chest.10-0608.
7. Overgaard D, Kaldan G, Marsaa K, Nielsen TL, Shaker SB, Egerod I. The lived experience with idiopathic pulmonary fibrosis: a qualitative study. Eur Respir J. 2016;47(5):1472–80. https://doi.org/10.1183/13993003.01566-2015.
8. Bausewein C, Farquhar M, Booth S, Gysels M, Higginson IJ. Measurement of breathlessness in advanced disease: a systematic review. Respir Med. 2007;101(3):399–410.
9. Bausewein C, Booth S, Gysels M, Kühnbach R, Haberland B, Higginson IJ. Understanding breathlessness: cross-sectional comparison of symptom burden and palliative care needs in chronic obstruc-

tive pulmonary disease and cancer. J Palliat Med. 2010;13(9):1109–18. https://doi.org/10.1089/jpm.2010.0068.

10. Funk GC, Kirchheinner K, Burghuber OC, Hartl S. BODE index versus GOLD classification for explaining anxious and depressive symptoms in patients with COPD—a cross sectional study. Res Rev. 2009;10(1):1. https://doi.org/10.1186/1465-9921-10-1.
11. Holland A, Fiore J Jr, Bell EC, Goh N, Westall G, Symons K, et al. Dyspnoea and comorbidity contribute to anxiety and depression in interstitial lung disease. Respirology. 2014;19(8):1215–21. https://doi.org/10.1111/resp.12360.
12. Ryerson CJ, Cayou C, Topp F, et al. Pulmonary rehabilitation improves long-term outcomes in interstitial lung disease: a prospective cohort study. Respir Med. 2014;108:203–10.
13. Bajwah S, Ross JR, Wells AU, et al. Palliative care for patients with advanced fibrotic lung disease: a randomised controlled phase II and feasibility trial of a community case conference intervention. Thorax. 2015;70:830–9.
14. Fulton BG, Ryerson CJ. Managing comorbidities in idiopathic pulmonary fibrosis. Int J Gen Med. 2015;8:309–18. https://doi.org/10.2147/IJGM.S74880.
15. Abu Youssefa HA, Sabryb Y, Sadekc M, Shabana M, Hafezd S. Study of the relationship of dyspnea with depression and functional status in patients with interstitial lung disease. Egypt J Chest Dis Tuberc. 2015;64(1):269–75. https://doi.org/10.1016/j.ejcdt.2014.11.019.
16. Van Manen M, Kreuter M, Van Den Blink B, Oltmanns U, Baroke E, Palmowski K, et al. What patients with pulmonary fibrosis and their partners think: a live, educative survey in the Netherlands and Germany. ERJ Open Res. 2017;3(1):00065-2016. https://doi.org/10.1183/23120541.00065-2016.
17. Lee YJ, Choi SM, Lee YJ, Cho Y-J, Yoon HI, Lee J-H, et al. Clinical impact of depression and anxiety in patients with idiopathic pulmonary fibrosis. PLoS One. 2017;12(9):e0184300. https://doi.org/10.1371/journal.pone.0184300.
18. Betancourt J, Torres del Castillo N, Hurtado H. Rehabilitación pulmonar en pacientes con enfermedad pulmonar intersticial difusa: estudio de casos. Rev Mov Cient. 2015;9(2):6–14.
19. Sgalla G, Cerri S, Ferrari R, Ricchieri M, Poletti S, Ori M, et al. Mindfulness-based stress reduction in patients with interstitial lung diseases: a pilot, single-centre observational study on safety and efficacy. BMJ Open Respir Res. 2015;2(1):e000065. https://doi.org/10.1136/bmjresp-2014-000065.
20. Swigris JJ, Brown KK, Make BJ, Wamboldt FS. Pulmonary rehabilitation in idiopathic pulmonary fibrosis: a call for continued investigation. Respir Med. 2008;102(12):1675–80.
21. The WHOQOL Group. Development of the World Health Organization WHOQOL-BREF quality of life assessment. Psychol Med. 1998;28:551–8.
22. Kaplan R, Bush J, Berry C. Health status: types of validity and the index of well-being. Health Serv Res. 1976;11(4):478–507.
23. Guyatt G, Berman L, Townsend M, Pugsley S, Chambers L. A measure of quality of life for clinical trials of chronic lung disease. Thorax. 1987;42(10):773–8.
24. Jones P, Quirk F, Baveystock C. The St. George's respiratory questionnaire. Respir Med. 1991;85:25–31.
25. Duck A, Spencer LG, Bailey S, Leonard C, Ormes J, Caress AL. Perceptions, experiences and needs of patients with idiopathic pulmonary fibrosis. J Adv Nurs. 2015;71(5):1055–65. https://doi.org/10.1111/jan.12587.
26. Sampson C, Gill BH, Harrison NK, Nelson AM, Byrne A. The care needs of patients with idiopathic pulmonary fibrosis and their carers (Canopy): results of a qualitative study. BMC Pulm Med. 2015;15:155. https://doi.org/10.1186/s12890-015-0145-5.
27. Molina J, Trigueros JA, Quintano JA, Mascarós E, Xaubet A, Ancochea J. Fibrosis pulmonar idiopática: un reto para la atención primaria. Idiopathic pulmonary fibrosis: a challenge for primary care. Semergen. 2014;40(3):134–42. https://doi.org/10.1016/j.semerg.2014.02.001.
28. Urrengoetxea LA, Solano CS, Barrenetxea MA, Moraza Cortés FJ, Quincoces AB, Sainz AC. Faseacelerada de la fibrosis pulmonar idiopática. Accelerated Phase of Idiopathic Pulmonary Fibrosis. Arch Bronconeumol. 2007;43(9):516–8.
29. Lindell KO, Liang Z, Hoffman LA, Rosenzweig MQ, Saul MI, Pilewski JM, et al. Palliative care and location of death in decedents with IPF. Chest. 2015;147(2):423–9. https://doi.org/10.1378/chest.14-1127.
30. Gainza D, Sanz EM, Alonso A, Varela M. Atención al final de la vida en los pacientes con fibrosis pulmonar idiopática. Med Paliat. 2017;24:21–5. https://doi.org/10.1016/j.medipa.2014.07.001.
31. Bajwah S, Higginson IJ, Ross JR, Wells AU, Birring SS, Riley J, et al. The palliative care needs for fibrotic interstitial lung disease: a qualitative study of patients, informal caregivers and health professionals. Palliat Med. 2013;27(9):869–76. https://doi.org/10.1177/0269216313497226.
32. Bradley B, Branley HM, Egan JJ, Greaves MS, Hansell DM, Harrison NK, et al. Interstitial lung disease guideline: the British Thoracic Society in collaboration with the Thoracic Society of Australia and New Zealand and the Irish Thoracic Society. Thorax. 2008;63 Suppl 5:v1–58. https://doi.org/10.1136/thx.2008.101691.
33. Kissane DW, Bobevski I, Gaitanis P, Brooker J, Michael N, Lethborg C, et al. Exploratory examination of the utility of demoralization as a diagnostic specifier for adjustment disorder and major depression. Gen Hosp Psychiatry. 2017;46:20–4. https://doi.org/10.1016/j.genhosppsych.2017.01.007.
34. van Manen MJ, Geelhoed JJ, Tak NC, Wijsenbeek MS. Optimizing quality of life in patients

with idiopathic pulmonary fibrosis. Ther Adv Respir Dis. 2017;11(3):157–69. https://doi.org/10.1177/1753465816686743.

35. Zigmond AS, Snaith RP. The hospital anxiety and depression scale. Acta Psychiatr Scand. 1983;67(6):361–70.
36. Quintana JM, Padierna A, Esteban C, Arostegui I, Bilbao A, Ruiz I. Evaluation of the psychometric characteristics of the Spanish version of the Hospital Anxiety and Depression Scale. Acta Psychiatr Scand. 2003;107(3):216–21.
37. Galiana L, Rudilla D, Oliver A, Barreto P. The Short Demoralization Scale (SDS): a new tool to appraise demoralization in palliative care patients. Palliat Support Care. 2017;9:1–8. https://doi.org/10.1017/S1478951516000973.
38. Rudilla D, Oliver A, Galiana L, Barreto P. A new measure of home care patients' dignity at the end of life: the Palliative Patients' Dignity Scale (PPDS). Palliat Support Care. 2016;14(2):99–108. https://doi.org/10.1017/S1478951515000747.
39. Benito E, Oliver A, Galiana L, et al. Development and validation of a new tool for the assessment and spiritual care of palliative care patients. J Pain Symptom Manage. 2014;47(6):1008–18.
40. Paul Victor CG, Treschuk JV. Critical literature review on the definition clarity of the concept of faith, religion, and spirituality. J Holist Nurs. 2019:898010119895368. https://doi.org/10.1177/0898010119895368.
41. Sinclair VG, Wallston KA. The development and psychometric evaluation of the Brief Resilient Coping Scale. Assessment. 2004;11:94–101.
42. Barreto P, Fombuena M, Diego R, Galiana L, Oliver A, Benito E. Bienestar emocional y espiritualidad al final de la vida. Med Paliat. 2015;22(1):25–32.
43. de la Revilla L, Bailón E, de Dios J, Delgado A, Prados M, Fleitas L. Validación de una escala de apoyo social funcional para su uso en la consulta del médico de familia. Atención Primaria. 1991;8(9):688–91.
44. Aaronson NK, Cull A, Kaasa S, Sprangers MAG. The EORTC modular approach to quality of life assessment in oncology. Int J Ment Health. 1994;23(2):75–96. https://doi.org/10.1016/0885-3924(95)94770-F.
45. Cohen J. Statistical power analysis for the behavioral sciences. Hillsdale: Lawrence Erlbaum; 1988.
46. Holland JC, Andersen B, Breitbart WS, et al. The NCCN distress management clinical practice guidelines in oncology. J Natl Compr Canc Netw. 2010;8:44884.
47. Julião M, Barbosa A, Oliveira F, Nunes B, Vaz Carneiro A. Efficacy of dignity therapy for depression and anxiety in terminally ill patients: early results of a randomized controlled trial. Palliat Support Care. 2013;11(6):481–9.
48. Rudilla D, Oliver A, Galiana L, Barreto P. Estudio comparativo de la eficacia del counselling y de la terapia de la dignidad en pacientes paliativos. Med Paliat. 2017;24:39–46. https://doi.org/10.1016/j.medipa.2014.09.005.
49. Levy MH, Back A, Benedetti C, Billings JA, Block S, Boston B, Bruera E, Dy S, Eberle C, Foley KM, Karver SB, Knight SJ, Misra S, Ritchie CS, Spiegel D, Sutton L, Urba S, Von Roenn JH, Weinstein SM. NCCN clinical practice guidelines in oncology: palliative care. J Natl Compr Canc Netw. 2009;7(4):436–73.
50. Houmann LJ, Rydahl-Hansen S, Chochinov HM, Kristjanson LJ, Groenvold M. Testing the feasibility of the Dignity Therapy interview: adaptation for the Danish culture. BMC Palliat Care. 2010;9:21.
51. Chochinov HM, Hassard T, McClement S, Hack T, Kristjanson LJ, Harlos M, et al. The patient dignity inventory: a novel way of measuring dignity-related distress in palliative care. J Pain Symptom Manage. 2008;36:559–71.
52. Chochinov HM, Hassard T, McClement S, et al. The landscape of distress in the terminally ill. J Pain Symptom Manage. 2009;38(5):641–9.
53. Chochinov HM, Kristjanson LJ, Breitbart W, et al. Effect of dignity therapy on distress and end-of-life experience in terminally ill patients: a randomised controlled trial. Lancet Oncol. 2011;12(8):753–62.
54. Albers G, Pasman HR, Rurup ML, de Vet HC, Onwuteaka-Philipsen BD. Analysis of the construct of dignity and content validity of the patient dignity inventory. Health Qual Life Outcomes. 2011;9:45.
55. Sanz A, Blancotoro L, Librada S, Julián-Caballero M, Del Valle M, Rocafort J. El concepto de calidad de vida en los profesionales de Cuidados Paliativos. Med Pal. 2008;15(3):165–70.
56. Rudilla D, Soto-Rubio A, Pérez MA, Galiana L, Fombuena M, Oliver A, et al. Psychological interventions in spirituality: a systematic review in palliative care. Med Paliativa (Internet). 2018;25(3):203–12. Available from: https://doi.org/10.1016/j.medipa.2016.10.004.
57. Puchalski CM, Vitillo R, Hull SK, Reller N. Improving the spiritual dimension of whole person care: reaching national and international consensus. J Palliat Med. 2014;17(6):642–56.