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Quality of life of idiopathic pulmonary fibrosis patients

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ABSTRACT: Little attention has been paid to quality of life (QOL) in idiopathic pulmonary fibrosis (IPF). Therefore, the aim of this study was to address this issue and study the relationship between QOL, depressive symptoms, and breathlessness in these patients.

Forty-one IPF patients and 41 healthy persons matched for age and sex completed the World Health Organization Quality of Life assessment instrument-100. The IPF patients also completed the Beck Depression Inventory, the Bath Breathlessness Scale, a social support questionnaire and a question concerning perceived seriousness of illness.

Compared to the control group, QOL in IPF patients was mainly impaired in the domains "physical health" and "level of independence". A number of relationships were found between pulmonary function tests and QOL. The QOL facet "negative feelings" was highly associated with scores on depression. Subjective breathlessness was related to depressive symptoms and QOL. Moreover, sex and effective/emotional breathlessness predicted overall QOL.

In conclusion, the impairment of the quality of life areas "physical health" and "level of independence" are important issues in idiopathic pulmonary fibrosis. Subjective breathlessness, especially the effective/emotional scale, seems related to quality of life and depressive symptoms. Rehabilitation programmes are needed that are aimed at physiological aspects and psychosocial aspects of idiopathic pulmonary fibrosis in order to enhance the quality of life of these patients.

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Idiopathic pulmonary fibrosis (IPF) or cryptogenic fibrosing alveolitis is a common interstitial lung disease of unknown aetiology. However, the clinical entity is well-defined. IPF is defined as "a specific form of chronic fibrosing interstitial pneumonia limited to the lung and associated with the histologic appearance of usual interstitial pneumonia (UIP) on surgical obtained lung biopsy" [1]. Most importantly, the diagnosis of IPF is made by exclusion of all known causes of fibrosing alveolitis [1, 2].

The patient demonstrates breathlessness, non-productive cough and bibasilar and inspiratory dry rates, and there are findings of finger clubbing. Constitutional symptoms such as weight loss, malaise, and fatigue may be present. Shortness of breath is usually the most salient and disabling symptom and is usually present at least 6 months before presentation [1].

The age at onset varies from 40–70 yrs. The clinical course is highly variable. However, in general, the prognosis is poor [1, 3–5]. The median survival after diagnosis is <5 yrs [1, 2, 5]. IPF is a potentially fatal disorder without spontaneous remission. Therefore, the majority of the patients require therapy. Subjective improvement occurs frequently. However, objective improvement in the decrease of symptoms, radiographic clearing, or physiological improvement, assessed by lung functions tests, occurs in <25% of treated patients [1, 2, 5, 6].

IPF patients have a limited life expectancy. In cancer patients, much is known about the impact of a short life expectancy. In a previous study [7], some IPF patients were shocked by the condition of other more severe IPF patients. This seems to indicate that patients are not fully aware that they have a fair chance of dying within a few years.

Regarding the quality of life (QOL) of IPF patients, little is known [1, 7]. Recently, a study among IPF patients with the aim of identifying the aspects of QOL that are relevant to this population was conducted. Results indicated that hobbies/leisure activities, mobility, transport, social relationships, working capacity, energy, and doing things slower were aspects relevant to IPF patients' QOL [7].

The severity of IPF, the poor prognosis and, in general, disappointing response to therapy resulting in a reduced life expectancy, emphasize the importance of this issue. Therefore, the aims of this study were to assess the QOL and depressive symptoms in IPF patients, the relationship between the QOL factors "fatigue" and "negative feelings". The study also aims to identify factors that are related to QOL (perceived illness severity, illness duration), and to identify impaired aspects of QOL to guide future rehabilitation programmes. Based on a previous study [7], QOL measurement was expanded with additional social support questions. Shortness of breath is a prominent

feature of IPF and it can have a major impact on patients' QOL [8]. Therefore, breathlessness was also measured with the aim of examining the relationship between QOL and objective (clinical data) as well as subjective breathlessness, and establishing the influence of (objective and subjective) breathlessness on overall QOL.

Material and methods

Subjects

Patients suffering from IPF from nine participating Dutch hospitals (Hospital Gelderse Vallei, Bennekom; Ignatius Hospital, Breda; Medical Centre Den Bosch, Den Bosch; St. Anna Hospital, Geldrop; Twee Steden Hospital, Tilburg; St. Radboud Hospital and Rehabilitation Centre Dekkerswald, Nijmegen; District Hospital Middle Twente, Hengelo; Maasland Hospital, Sittard; and University Hospital, Maastricht) were selected. The diagnosis of IPF was based on consistent clinical features, together with radiographical findings, bronchoalveolar lavage fluid analysis results and finally, biopsy proven with histological characteristics of UIP consistent with IPF [1, 3].

In the context of two other studies [9], 555 persons had indicated their willingness to participate in a study on QOL. From the group of participants who returned a completed test-booklet (n=356; 64.1%), matched healthy control subjects (n=41) were selected on the basis of sex and age (table 1). These control subjects were used in order to compare their World Health Organization Quality of Life assessment instrument-100 (WHOQOL-100) scores with those of the IPF subjects.

Measures

All subjects completed the WHOQOL-100 (Dutch version [10]), a cross-culturally developed, generic, multidimensional QOL measure [11, 12]. It consists of 100 items assessing 24 facets of QOL within six domains (physical health, psychological health, level of independence, social relationships, environment, and spirituality/religion/personal beliefs) and a general evaluative facet (overall QOL and general health) [1]. Each facet is represented by four items. The response scale is a 5-point Likert scale. Scores on each facet and domain can range from 4–20. With the exception of the facets "pain and discomfort", "negative feelings", and "dependence on medication or treatments", where low scores indicate higher QOL, higher scores indicate a better QOL. The reliability and validity of the instrument, which has also been tested in sarcoidosis, are good [13–15]. The test-retest reliability is satisfactory [9].

Patients also completed the Beck Depression Inventory (BDI) [16], a well-validated index of depression which correlates well with diagnostic criteria. This instrument consists of statements which are arranged in 21 groups of four possible responses. Patients were asked to select one statement from each group that best described the way they felt the previous week. Each answer is scored on a four-point Likert-type scale from 0–3. A summation of the ratings in the 21 groups indicates the severity of depression (possible range 0–63). A patient with a total score of 15 or above was considered to have significant depressive symptoms. In order to control the physical effects of the illness on mood, items referring to physical symptoms were excluded. The scale based on the remaining 15 items was entitled the Cognitive Depression Inventory (CDI). This appears to be a valid way to prevent confounding relationships [17].

Patients also completed the Bath Breathlessness Scale (BBS) [18]. This is a 35-item adjective subjective breathlessness scale measuring four aspects of breathlessness: physical sensations, effective/evaluative descriptions, low energy items, and hyperventilation/speechless items. In addition, patients were asked to indicate on an 8-point response scale how breathless they had been during the last 2 weeks and were asked whether their breathlessness is brief, periodic or continuous. Apart from the four separate scale scores and the severity score, a total BBS score can be calculated. Patients with a respiratory, oncology or cardiac diagnosis had provided, in interviews, 63 descriptions of breathlessness. Subsequently, expert health professionals sorted these descriptions according to similarity and judged them on frequency and

Table 1. – Characteristics of the idiopathic pulmonary fibrosis (IPF) patient population and the control group

	IPF group	Control group
Demographic characteristics		
Number of cases n	41	41
Sex male/female	15/26	15/26
Age yrs	63.5 (38–80)	64.0 (45–83)
Smoking yes/no/missing	4/35/6	
Medical characteristics		
Oxygen use yes/no	14/27	
If yes now often? 24 h a day/sometimes	6/8	
Steroid use at time of study yes/no	28/13	
Time since diagnosis yrs	5.8±5.4	
FEV_1 % pred	70.1±21.5	
$T_{L,CO}$ % pred	45.3±14.9	
P_{O_2} at rest kPa	9.1±1.7	
P_{O_2} during exercise kPa	7.0±1.2	
P_{O_2} difference score kPa	-2.7±1.5	
Symptoms		
Cough yes/no	35/6	
Reduced exercise capacity yes/no	34/7	
Severity of dyspnoea, possible range 0–7	3.8±2.0	
Psychological characteristics		
BDI-scores	11.9±6.0	
CDI-scores	7.0±5.1	

Data are expressed in means, (range) or as mean±SD. FEV_1: forced expiration volume in one second; $T_{L,CO}$: transfer factor of the lung for carbon monoxide; P_{O_2}: arterial oxygen tension; BDI: Beck Depression Inventory; CDI: Cognitive Depression Index.

severity. The final structure was established using multivariate analyses [18].

Patients completed 14 questions which were specifically developed for chronically ill persons [19]. These questions refer to three different kinds of social support: practical support ("Do you get practical support from your children?"), emotional support ("Do you get emotional support from your partner?"), and understanding ("Do you get understanding from your children?"). Each type of social support question was asked in relation to a set of possible support providers such as the patient's partner, children and neighbours. A total social support score is calculated by summing-up the general questions of the three types of support. In addition, on a 4-point rating scale (not at all serious, not serious, serious, very serious) patients were asked how serious they thought their IPF was. Finally, patients' physicians provided information concerning illness duration and patients provided information concerning symptoms.

Assessment procedures

Lung function measures included forced expiratory volume in one second (FEV1) and inspiratory vital capacity measured with a pneumotachograph. Measures were selected from the three best efforts. The transfer factor of the lung for carbon monoxide ($T_{L,CO}$) was measured by the single breath method (Masterlab; Jaeger, Würzburg, Germany). Values were expressed as a percentage of those predicted [20].

Exercise capacity

Patients performed a symptom limited incremental exercise test on an electronically braked cycle ergo meter (Cornival 400, Lode, Groningen, the Netherlands). After a period of rest and 2 min of unloaded pedalling, a progressively increasing work rate test was started in order to determine peak work rate and oxygen uptake of the subject. The work rate increase was set at $10\ W \cdot min^{-1}$ for each patient and $15-30\ W \cdot min^{-1}$ for the healthy volunteers, depending upon their training status. Pedalling frequency was selected by the subjects (between 60–70 rpm) and held constant throughout the test. Breath-by-breath gas exchange was measured throughout the test by a ventilated hood system (Oxyconbeta, Jaeger, Bunnik, the Netherlands). An infrared electrode was placed on a finger to measure oxygen saturation (Fasttrac, Sensor Medics Co., Anaheim, CA, USA). Heart rate was measured throughout the test using a Sport tester (PE3000, Polar Electro cy, Kempele, Finland). At rest and during maximum exercise, arterial blood samples were taken and arterial blood gas analyses were performed.

Statistical procedure

Data are expressed as mean ± SD unless otherwise stated. Within the IPF group, t-tests were used for the following comparisons: oxygen use *versus* no oxygen use for QOL; steroid use *versus* no steroid use for QOL and dyspnoea. Furthermore, t-tests were used to test for differences between the IPF and healthy groups. A p-value of <0.005 was considered to be statistically significant. In the IPF group Pearson, or where appropriate, Spearman correlations were used to study: 1) the association between FEV1, $T_{L,CO}$, arterial oxygen tension (P_{a,O_2}) at rest, P_{a,O_2} after exertion, P_{a,O_2} difference and illness duration *versus* the QOL (including the additional social support questions) and seriousness of the illness; 2) the association between subjective breathlessness *versus* QOL; 3) the relationship between seriousness of IPF *versus* QOL and the BDI/CDI; and 4) the association between the WHOQOL-100 facets "negative feelings" and "energy and fatigue" *versus* the BDI/CDI. Stepwise multiple regression was performed to establish the influence of (objective and subjective) breathlessness on overall QOL. A p-value <0.05 was considered statistically significant. All analyses were performed using the Statistical Package for Social Sciences (SPSS) for Windows (SPSS, Chicago, IL, USA).

Results

The demographic (also healthy group), medical and psychological characteristics, as well as complaints of the IPF group are summarized in table 1. The comparison of QOL scores between the IPF group and the control group are shown in table 2. It appeared that the QOL of the IPF group was impaired with regard to their general QOL and the domains "physical health" and "level of independence". At facet level, IPF patients had problems with "pain and discomfort", "energy and fatigue", "sleep and rest", "activities of daily living", "dependence on medication or treatments", "working capacity", "ability to acquire new information and skills", and "participating in, and possibilities for recreation/leisure". The QOL of IPF patients using oxygen did not differ from IPF patients who did not use oxygen. Steroid use was unrelated to dyspnoea and QOL.

Patients' FEV1 appeared to be related to the domains "psychological health", "social relationships", and "environment". Furthermore, FEV1 was significantly correlated with "overall QOL", "pain and discomfort", "positive feelings", "self-esteem", "negative feelings", and "participating in, and possibilities for recreation/leisure" (table 3). Arterial oxygen saturation (S_{a,O_2}) decrease on exertion was related to the QOL domain "level of independence" and the facet "transport and P_{a,O_2} at rest" was positively correlated with patients' "overall QOL". The $T_{L,CO}$ was not associated with any QOL aspect (table 3). No relationship was found between subjective breathlessness and the question on perceived seriousness of illness, on the one hand, and lung function parameters or illness duration, on the other hand. With regard to the social support questionnaire, emotional support was negatively related to FEV1, whereas "understanding" and the total social support score were negatively related to S_{a,O_2} decrease on exertion. Illness duration was

Table 2. – Quality of life (QOL) scores measured with the World Health Organization Quality of Life assessment instrument-100 (WHOQOL-100) of the idiopathic pulmonary fibrosis (IPF) patient population compared to the matched control group

	IPF group	Control group	T-value
Overall QOL and general health	**13.1±3.0**	**16.5±2.1**	**5.98****
Physical health	**12.7±2.3**	**15.9±2.2**	**6.03****
Pain and discomfort	11.4±2.9	8.5±3.1	-3.81**
Energy and fatigue	10.6±2.9	15.1±2.9	6.61**
Sleep and rest	14.5±3.5	16.9±3.4	3.09^f
Psychological health	**13.8±2.4**	**15.1±2.0**	NS
Self-esteem	13.0±2.6	15.1±3.1	3.15^f
Level of independence	**12.0±2.9**	**17.2±2.0**	**8.78****
Mobility	12.1±3.3	16.8±2.8	6.80**
Activities of daily living	12.5±3.6	17.1±2.0	6.66**
Dependence on medication or treatments	12.0±4.0	6.0±2.2	-7.60**
Working capacity	10.6±3.9	16.7±2.9	7.56**
Environment	**15.1±1.8**	**16.0±1.9**	NS
Ability to acquire new information and skills	14.2±2.2	16.1±2.6	3.34^f
Participating in and possibilities for recreaction/leisure	13.6±3.3	16.1±2.9	3.53^f

Data are expressed as mean±SD. Domains are in bold type and facets belonging to these domains are listed underneath. Nonsignificant facets have not been included in the above table. Higher scores on QOL facets and domains indicate better QOL. Exceptions are the facets 'pain and discomfort', 'negative feelings', and 'dependence on medication or treatments'. ^{f}: $p<0.005$; **: $p<0.001$; NS: nonsignificant.

associated with "ability to acquire new information and skills" (table 3).

Perceived seriousness of illness was related to the QOL domains "physical health" ($r=-0.47$; $p<0.05$) and "level of independence" ($r=-0.44$; $p<0.05$) and the QOL facets "pain and discomfort" ($r=0.49$; $p<0.05$), "energy and fatigue" ($r=-0.64$; $p<0.005$), "dependence on medication or treatments" ($r=-0.50$; $p<0.05$), "working capacity" ($r=-0.57$; $p<0.01$), and "sexual activity" ($r=0.66$; $p<0.005$). Seriousness of illness appeared to be unrelated to depressive mood (BDI/CDI).

The number of patients with a BDI score of $\geqslant 15$ was eight (23.5%; $n=34$), whereas nine patients (26.5%; $n=34$) had a CDI score of $\geqslant 9$. Both BDI and CDI scores correlated highly with the WHOQOL-100 facet score "negative feelings" ($r=0.73$ and $r=0.75$; $p<0.001$, respectively). The correlations between the BDI and CDI scores, and the WHOQOL-100 "energy and fatigue" facet scores were not significant. The BDI was related to the breathlessness scale "affective/emotional" ($r=0.55$; $p<0.01$) whereas the CDI was correlated with the breathlessness scales "affective/emotional" ($r=0.53$; $p<0.01$) and "low energy" ($r=0.44$; $p<0.05$).

The correlations between subjective breathlessness

Table 3. – Significant correlations between World Health Organization Quality of Life assessment instrument-100 (WHOQOL-100) of the idiopathic pulmonary fibrosis (IPF) patient population and forced expiration volume in one second (FEV_1), some arterial blood gas analysis results and illness duration

	FEV_1	Arterial oxygen desaturation	P_{O_2} at rest	Illness duration
Overall QOL and general health	0.36		0.41	
Physical health				
Pain and discomfort	-0.41			
Psychological health	**0.39**			
Positive feelings	0.42			
Self-esteem	0.34			
Negative feelings	-0.40			
Level of independence		**0.55**		
Social relationships	**0.38**			
Environment	**0.33**			
Ability to acquire new information and skills				0.40
Participating in and possibilities for recreaction/leisure	0.34			
Transport		0.55		
Emotional support	-0.52			
Understanding support		-0.55		
Total social support		-0.60		

All correlations are significant at $p<0.05$ level. Higher scores on quality of life (QOL) facets and domains indicate better QOL. Exceptions are the facets 'pain and discomfort' and 'negative feelings'. P_{O_2}: arterial oxygen tension.

Table 4. – Significant correlations between World Health Organization Quality of Life assessment instrument-100 (WHOQOL) scores of the idiopathic pulmonary fibrosis (IPF) patient and population subjective dyspnoea (Bath Breathlessness Scale (BBS)) scores

	Severity[#]	I	II	III	IV	Total
Overall QOL and general health				-0.55[f]		-0.56[§]
Physical health			**-0.38**[§]	**-0.59**[f]		**-0.48**[§]
Pain and discomfort				0.52**		
Energy and fatigue	-0.57	-0.46[§]	-0.54[f]	-0.74[f]		-0.69[f]
Psychological health			**-0.45**[§]	**-0.66**[f]		**-0.63**[f]
Positive feelings			-0.44[§]	-0.52[f]		-0.55[§]
Cognitive functions[¶]				-0.53[f]	-0.46[§]	-0.52[§]
Self-esteem				-0.49[f]		
Bodily image and appearance			-0.43[§]	-0.44[§]		
Negative feelings			0.56[f]	0.68**		0.68[f]
Level of independence			**-0.45**[§]	**-0.41**[§]		
Activities of daily living				-0.49[§]		
Working capacity				-0.56[f]		
Social relationships			**-0.42**[§]	**-0.47**[f]		
Personal relationships				-0.54[f]		-0.56[§]
Social support			-0.40[§]	-0.42[§]		
Sexual activity		0.45[§]				
Environment			**-0.41**[§]	**-0.54**[f]		**-0.72**[§]
Physical safety and security						-0.48[§]
Home environment				-0.44[§]		-0.52[§]
Financial resources				-0.48[f]		-0.56[§]
Participating in and possibilities for recreaction/leisure			-0.43[§]	-0.50[f]		-0.52[§]
Physical environment[+]		-0.46[§]	-0.54[f]	-0.45[§]		-0.74**

Data presented are correlation coefficients. Bold text represents domains with facets listed underneath; nonsignificant facets have not been included in the table. [#]: I: BBS physical sensations; II: BBS Affective/evaluative; III: BBS Low energy; IV: BBS Hyperventilation/speechless; Total: total BBS score; [¶]: Thinking, learning, memory, and concentration; [+]: pollution/noise/traffic/climate. Higher scores on QOL facets and domains indicate better QOL. Exceptions are the facets 'pain and discomfort' and 'negative feelings'. [§]: $p<0.05$; [f]: $p<0.01$; **: $p<0.001$.

and QOL are presented in table 4. As can be seen, shortness of breath is related to most domains and facets of patients' QOL. Exceptions are the QOL facets "sleep and rest", "mobility", "dependence on medication", "health and social care", "ability to acquire new information and skills", and "transport" as well as the domain "spirituality/religion/personal beliefs" which showed no significant correlation with any of the BBS scales or the total BBS score.

Subsequently, a stepwise multiple regression analysis was performed to examine the relationship between breathlessness and QOL further. A conservative strategy was adopted in which the demographic variables age and sex were entered as a block in the first step of the regression analysis and the pulmonary function parameters (FEV1, $T_{L,CO}$, and P_{a,O_2} difference score) as a block in the second step of the analysis to statistically adjust for their influence, despite negligible associations with overall QOL and breathlessness. In the final model with overall QOL as dependent variable ($R^2=0.98$; $F(6,2)=20.8$, $p<0.05$), sex ($\beta=-0.74$; $t=-5.3$, $p<0.05$) and effective/emotional descriptions of breathlessness ($\beta=-1.16$; $t=-7.3$; $p<0.05$) were the only factors that significantly contributed to the prediction of overall QOL.

Age and sex were not associated with depression (BDI and CDI), negative feelings, duration of illness, FEV1, $T_{L,CO}$, and S_{a,O_2} decrease on exertion. Age was related to both perceived seriousness of illness ($r=-0.44$; $p<0.05$; $n=21$) and the BBS scale "physical sensations" ($r=-0.38$; $p<0.05$), whereas sex was not.

Discussion

The QOL of the IPF patients was mainly impaired in the domains "physical health" and "level of independence" compared with a matched healthy control group. In general, these results are similar to those found in another common interstitial lung disease, *i.e.* sarcoidosis [13].

In contrast to previous findings in sarcoidosis [13], the FEV1 appeared to be related to QOL. Especially, "psychological health" (domain level), "social relationships" (domain level), "environment" (domain level), "overall QOL", "pain", "positive feelings", "negative feelings", "self-esteem", and "recreation/leisure" appeared to be related to the FEV1. When comparing the aspects of QOL associated with FEV1, with the facets in which the QOL of IPF patients is impaired, it appears that the results are only congruent for "overall QOL", "pain", "self-esteem", and "recreation/leisure". It is striking that the QOL domains impaired in IPF patients are not associated with FEV1. Moreover, the other lung function parameters were hardly related to patients' QOL. An exception was S_{a,O_2} decrease during exertion, which was related to the QOL domain "level of independence" and the facet "transport". Although caution in the interpretation is warranted, as the sample size of the studied patient population was rather small, it is reasonable to assume that although FEV1 was related to QOL, lung function parameters, except S_{a,O_2} decrease during exertion, are not reliable indicators of QOL impairment in IPF patients.

Subjective dyspnoea, especially the effective/evaluative and low energy scales, appeared to be correlated with many domains and various aspects of QOL. However, subjective dyspnoea was not related to the pulmonary function parameters which are measures of lung capacity and thus, indirectly, objective dyspnoea measures. Patients′ perception of dyspnoea is not associated with objective breathlessness measures. The regression analysis showed that the male IPF patients and patients with a low score on effective/emotional breathlessness indicated a better overall QOL.

The major QOL problems of the IPF patients appeared to be fatigue, mobility, activities of daily living, and working capacity. As mentioned above, none of these QOL aspects was significantly related to the lung function parameters. Although abnormal physical exhaustion and fatigue are often regarded as a natural consequence of pulmonary diseases, in line with the present observations, GUGGER [21] assumed that, apart from factors specifically related to pulmonary diseases, psychosocial factors are mainly responsible for the impaired physical fitness and fatigue. Therefore, it was recommended that a careful evaluation of psychological aspects be included in the diagnostic evaluation of patients suffering from a certain pulmonary disorder [21]. Moreover, the assessment, support, and reinforcement of a patient′s psychosocial assets and ability to cope with chronic respiratory difficulty can help enhance the QOL, and decrease feelings of fear for the manifestations and/or treatments of disease [22–24].

The patients′ perception regarding the severity of their illness was not related to objective measures of severity and illness duration. This indicates that the patients′ own opinions about the severity of their disease is not in accordance with objective data. This is in alignment with the fact that patients frequently report subjective improvement despite no objective improvement [1, 2, 5, 6]. Possible explanations are that: 1) patients suppress information concerning the seriousness of their disease; 2) patients have psychologically adapted to their situation; and/or: 3) patients feel that their situation is better than indicated by their physicians. Which of these reasons play a key role, needs to be explored.

Perceiving IPF as a severe disease is related to impaired physical health, as well as reduced feelings of independence from others. More specifically, disease severity is associated with more pain, more fatigue, an impaired working capacity and more dependency on medication. Thus, the perceived seriousness of IPF has a negative influence on patients′ QOL. Surprisingly, perceiving IPF as a severe illness was related to more sexual activity. Presumably, patients want to enjoy life as much as possible and, therefore, take advantage of their sexual activity, in one of its many forms, as long as possible. Studies among populations of rheumatoid arthritis, psoriasis, and sarcoidosis patients [25, 26] revealed no sexual difficulties.

Remarkably, the perceived seriousness of IPF, as well as fatigue scores, were not related to depressed mood. In a study among sarcoidosis patients, a relationship was found between fatigue and depression scores [13]. The authors realize that the sample size was rather small. Therefore, these results should be interpreted with care. A larger study is needed in order to replicate the results from the present study. However, considering the high correlation between the BDI and the WHOQOL-100 facet "negative feelings", using a separate measure for assessing depressive mood seems to be redundant. Further studies exploring this issue are required.

In the present study, 23.5% of the patients who fully completed the BDI had a score indicative of significant depressive symptoms. Using the CDI score, which excludes physical symptoms of depression, comparable results emerged. In another study, IPF patients who participated in focus groups spontaneously mentioned feelings of depression [7]. It emerges that more attention should be paid to the negative feelings of IPF patients. The fact that general fatigue (measured with the WHOQOL), a symptom of clinical depression, was unrelated to the BDI/CDI scores implies that although IPF patients are bothered by negative feelings, they are in general, not clinically depressed. The relationship between the CDI and the breathlessness scale "low energy" (physical fatigue) might indicate that dyspnoea causes fatigue. Previously, it appeared, that in a group of sarcoidosis patients [13], the presence of symptoms played a major role regarding those feelings. Moreover, sarcoidosis patients with complaints demonstrated higher BDI scores compared to patients without complaints [27].

Most of the aspects of impaired QOL in IPF are not measured by disease-specific health status (or nowadays also called health-related QOL) lung questionnaires such as the St. George′s Respiratory Questionnaire (SGRQ). QOL has a much wider scope than the physical, emotional, and social domains usually studied in health status measures. Health status measures assess fewer aspects and, therefore, provide less specific information concerning patients′ problems. Moreover, aspects such as fatigue are not usually assessed in health status measures. Furthermore, QOL instruments measure a person′s own evaluation of their functioning, whereas health status measures assess patients′ physical, emotional and social functioning [7, 28]. The reliability and validity of the WHOQOL-100 are high [14]. Within a group of sarcoidosis patients the WHOQOL-100 distinguishes well between patients with and without complaints [13, 15]. Previously, it was established that the WHOQOL-100 was preferable to the SGRQ in IPF [7]. Reasons were, for instance, that focus group participants made substantial negative remarks about the SGRQ, the scales of this questionnaire were not mentioned as important aspects of life and the SGRQ only measures three aspects (symptoms, activity, and impact). This provides very limited information with regard to problems that patients may experience [7]. In line with this, results from the present study indicate that the WHOQOL-100 appeared to be useful in IPF. In contrast, CHANG *et al.* [29] studied a group of interstitial lung disease patients (33 IPF, 10 sarcoidosis, seven miscellaneous) and concluded that the Short Form 36 Item Questionnaire (SF-36) and SGRQ are good tools for measuring health-related QOL or health status. The discrepancy between their findings and

other previous findings [7] might be caused by the fact that focus groups were used in other studies, in which IPF patients themselves could comment on the measures and did not rely solely on psychometrics. The SF-36 might be a good measure for assessing health status in IPF. The choice between a health status measure and a QOL measure should be based on the aim of a study [28]. If the interest is only in functioning, then the SF-36 might be suitable [29, 30]. However, if the interest is in the opinion of the patients, then the WHOQOL-100 is a suitable measure. Before using the SF-36, the usefulness and acceptability of the instrument should be studied from the patients′ view point with, for example, focus groups or individual interviews.

Although the severity, disablement and shortened life expectancy of IPF might be comparable with another serious illness such as cancer, less attention has been paid to these issues and until now hardly any rehabilitation programmes have been prepared for patients suffering from IPF. This study intended to focus on the aspects of QOL impairment of IPF patients. Rehabilitation programmes should be developed to improve the knowledge of patients and their families about the severity of the disease and to improve adaptive coping with the disease. In a study among chronic leukaemia patients, coping reflected the different strategies adopted to manage patients′ capacity for getting through difficult times. The adaptive strategy action included acquiring as much information as possible about their disease [31].

In conclusion, the quality of life of idiopathic pulmonary fibrosis patients was mainly impaired in the areas "physical health" and "level of independence". Perceived seriousness of idiopathic pulmonary fibrosis was related to the same quality of life domains. The World Health Organization Quality of Life assessment instrument-100 facet "negative feelings" appeared to be useful to assess depressive mood. Fatigue was unrelated to the depressive mood. Furthermore, although quality of life aspects were related to lung function impairment, it only concerned nonimpaired aspects. Subjective breathlessness was associated with depressive mood and quality of life. Finally, sex and the effective/emotional breathlessness scale were the only predictors of overall quality of life. Therefore, it is reasonable to assume that the assessment, support and reinforcement of a patient′s psychosocial assets and ability to cope with idiopathic pulmonary fibrosis can help enhance the quality of life. Moreover, these aspects should be a primary consideration in the development of rehabilitation programmes for patients suffering from idiopathic pulmonary fibrosis as well as pathophysiological aspects of this severe pulmonary disorder.

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