Tilburg University

Quality of life and health status in interstitial lung diseases

de Vries, J.; Drent, M.

Published in:
Current Opinion in Pulmonary Medicine

Publication date:
2006

Document Version
Publisher's PDF, also known as Version of record

[Link to publication in Tilburg University Research Portal](#)

Citation for published version (APA):
de Vries, J., & Drent, M. (2006). Quality of life and health status in interstitial lung diseases. *Current Opinion in Pulmonary Medicine*, 12(5), 354-358.

General rights

Copyright and moral rights for the publications made accessible in the public portal are retained by the authors and/or other copyright owners and it is a condition of accessing publications that users recognise and abide by the legal requirements associated with these rights.

- Users may download and print one copy of any publication from the public portal for the purpose of private study or research.
- You may not further distribute the material or use it for any profit-making activity or commercial gain
- You may freely distribute the URL identifying the publication in the public portal

Take down policy

If you believe that this document breaches copyright please contact us providing details, and we will remove access to the work immediately and investigate your claim.

Quality of life and health status in interstitial lung diseases

Jolanda De Vries^a and Marjolein Drent^b

Purpose of review

The aim of this review is to provide information on the influence of interstitial lung disease on patients' health status and quality of life, with an emphasis on which aspects might be affected by interstitial lung disease.

Recent findings

The quality of life of sarcoidosis patients with current symptoms and patients with idiopathic pulmonary fibrosis is mainly impaired in the domains of physical health and level of independence. Concerning health status, sarcoidosis patients report reduced physical functioning, especially when they have current symptoms. The health status of idiopathic pulmonary fibrosis patients is impaired in almost all aspects. Subjectively assessed dyspnea is related to health status. A relationship between treatment and health status has not been found.

Summary

Results from the existing studies show that both illnesses have a substantial impact on patients' life. Studies aiming at measuring quality of life or health status in other interstitial lung diseases are scarce or nonexistent.

Keywords

health status, idiopathic pulmonary fibrosis, interstitial lung disease, quality of life, sarcoidosis

Curr Opin Pulm Med 12:354–358. © 2006 Lippincott Williams & Wilkins.

^aDepartment of Psychology and Health, Medical Psychology, Tilburg University and St Elisabeth Hospital, Tilburg and ^bDepartment of Pulmonology, University Hospital Maastricht, The Netherlands

Correspondence to J. De Vries, Tilburg University, Department of Psychology and Health, Medical Psychology, PO Box 90153, 5000 LE Tilburg, The Netherlands
Tel: +31 134662167; e-mail: j.devries@uvtnl, Jvries@elisabeth.nl

Current Opinion in Pulmonary Medicine 2006, 12:354–358

Abbreviations

ILD interstitial lung disease
IPF idiopathic pulmonary fibrosis
QOL quality of life

© 2006 Lippincott Williams & Wilkins
1070-5287

Introduction

The term interstitial lung disease (ILD) embraces a wide variety of diseases that are characterized by inflammation, damage and fibrosis in the acinar regions of the lungs. The most common ILD is sarcoidosis, followed by idiopathic pulmonary fibrosis (IPF). Other ILDs are, among many others, more or less orphan diseases Wegener's granulomatosis, pneumoconiosis and bronchiolitis obliterans syndrome.

Sarcoidosis is a disease of young persons with a peak between 20 and 40 years [1,2]. The clinical course of sarcoidosis is highly variable and almost every organ can be involved. The lungs are affected in more than 90% of sarcoidosis patients. Furthermore, the lymph nodes, skin and eyes are frequently involved. Patients with pulmonary sarcoidosis may present with symptoms related directly to the chest such as coughing, dyspnea on exertion, chest pain, chest discomfort and wheezing. Other symptoms such as fever, anorexia, weight loss, general weakness and pain are also related to sarcoidosis [3,4]. A number of studies showed that fatigue is a prominent feature of sarcoidosis [4–7].

IPF, also known as cryptogenic fibrosing alveolitis, is a chronic, progressive, usually fatal disease for which no adequate pharmacotherapy is available. The disease is characterized by inflammation and fibrosis of the alveolar walls [8]. Mostly, corticosteroids are used in combination with azathioprine or cyclophosphamide [9]. Patients with end-stage IPF may be candidates for lung transplantation. The most important symptoms are dyspnea on exertion, fatigue and cough [10–13].

In general, fatigue, dyspnea and cough are the main symptoms in ILD [14]. These symptoms are disabling for the patient and cause an impaired quality of life (QOL). The aim of this review is to provide information on the influence of ILD on patients' health status and QOL, with an emphasis on which aspects might be affected by ILD.

Methods

A search using the PubMed database was performed with the keywords 'interstitial lung disease and health status' and 'interstitial lung disease and quality of life'. This resulted in 77 and 112 articles, respectively. From these 189 articles, 150 were dropped based on the title or abstract. Reasons for excluding articles were language, i.e. not written in English, German or Dutch ($n=28$);

patients were children ($n=10$); studies with animals ($n=2$); appeared in both searches ($n=35$); head and neck Wegener ($n=2$); occupational diseases ($n=2$); studies concerning ILD as a result of lung cancer ($n=5$); studies on lung transplantation ($n=20$) or bronchiolitis obliterans syndrome ($n=3$); and health status or QOL were not examined by means of questionnaires in one or more ILDs ($n=56$).

Definitions of quality of life and health status

QOL and health status are two different concepts. Health status refers to the impact of disease on patients' physical, psychological and social functioning [15], while QOL refers to patients' perception or evaluation of their functioning [10,15,16]. The WHOQOL Group defined QOL as 'a person's perception of their position in life within the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns. It is a broad-ranging concept incorporating, in a complex way, a person's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to salient features of the environment' [17]. This definition reflects the view that QOL refers to a subjective evaluation of functioning, which is embedded in a cultural, social and environmental context. Furthermore, QOL is a very broad multidimensional concept, going beyond the World Health Organization's definition of health [18]. Health-related QOL is QOL, but restricted to the physical, mental and social domain. In addition health status measures are subjective in the sense that they are completed by patients themselves. QOL measures are not only completed by patients, but also ask to evaluate the aspects of QOL (e.g. 'How satisfied...?', 'How bothered...?') [19]. Thus, health status and (health-related) QOL are distinct concepts [16].

Although most of the studies suggest they measure QOL, they actually assess health status. The studies discussed in the present review are divided into QOL and health status studies.

Outcome of quality of life studies

Research on QOL in patients with ILD has existed for less than 10 years, and has only been done among sarcoidosis and IPF patients.

Sarcoidosis

The greatest impact on QOL in sarcoidosis, as seen in clinical practice, is caused by rather nonspecific symptoms that are hard to objectify such as fatigue and sleeping disorders [7]. For instance, in one study [6], there appeared to be a number of QOL areas in which patients, particularly those with current symptoms, experienced problems. Surprisingly, sarcoidosis patients either with or without current symptoms suffered from

fatigue, sleeping problems and impaired general QOL compared with a healthy control group. Apart from the physical problems mentioned above, patients with current symptoms suffered from impaired QOL with regard to their mobility, working capacity and activities of daily living. Thus, in agreement with other studies [20–22], sarcoidosis has a considerable impact on patients' QOL, especially in those patients with current symptoms.

In one of these latter studies [22], the QOL of sarcoidosis patients was compared with the QOL of rheumatoid arthritis patients and healthy controls. Compared with the QOL of the healthy controls, the QOL of both patient groups was impaired with regard to the domains of physical health and level of independence. The rheumatoid arthritis group scored even lower than the sarcoidosis group on overall QOL and health. Fatigue, sleep, activities of daily living and working capacity were major problems in sarcoidosis as well as rheumatoid arthritis patients. To date, rheumatoid arthritis patients have demonstrated more problems related to pain and mobility [22]. Another study focusing on gender differences revealed that female patients had a lower QOL, except for the aspect of positive feelings [21].

Idiopathic pulmonary fibrosis

Two studies [11,23] have been performed establishing QOL in IPF patients. One [23] concerned a semiquantitative study in which 10 patients participated in focus groups about the face validity of a QOL and a health status measure. In addition, patients talked about major issues relating to their QOL. Hobbies/leisure activities, mobility, transport, social relationships, working capacity, energy and doing things slower were aspects relevant to IPF patients' QOL. Furthermore, compared with the St George Respiratory Questionnaire – a chronic obstructive pulmonary disease-specific health status measure – the WHOQOL-100 – a QOL questionnaire, with an additional social support questionnaire – appeared to be preferable [23]. The purpose of the other study was to examine the relationship between QOL, depressive symptoms and breathlessness in IPF patients. Compared with the control group matched for age and sex, QOL in IPF patients was mainly impaired in the domains of 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 [11].

Outcome of health status studies

With regard to health status, substantially more studies have been performed. Studies mainly focused on sarcoidosis or IPF. One study [24] concerned Wegener's

granulomatosis. This study was conducted among 60 patients, including 35 patients with manifestations in the lungs, aged below 40 years at diagnosis. The focus of the study was on unemployment. Compared with working patients, the unemployed had a worse health status. This was independent of disease severity and extent of disease. The only study [25] performed in patients with pneumoconiosis examined the relationship between alcohol consumption and health status in a group of 300 patients. Patients who had used alcohol during the previous year were younger, had less comorbidity and a better health status compared with patients who did not consume any alcohol. No studies were found with regard to bronchiolitis obliterans syndrome.

Interstitial lung disease

In a study [26] that aimed to assess the relationship between health status and physiological impairment, patients with various ILDs appeared to have a moderately reduced health status. As might be expected, the scores on the applied questionnaires were related to the dyspnea scores of patients and results on their pulmonary function tests.

Sarcoidosis

In two studies [6,27], patients scored higher on cognitive behavior, home management tasks, recreation and hobbies, sleep, social interaction, and work compared with a control group. In the study by Drent *et al.* [6], sarcoidosis patients suffering from symptoms appeared to be responsible for the differences between the sarcoidosis patients and the control group. Patients with current symptoms reported more depressive symptoms compared with patients without current symptoms. Moreover, whereas the latter subgroup experienced more positive affect, no difference between the two sarcoidosis subgroups were found with regard to negative affect. From the health status aspects, sleep appeared to be associated with depressive symptoms, in general, and depressive cognitions, in particular [6]. Cox *et al.* [28] found that higher scores on depressive symptoms and perceived stress were related to lower health status scores. In another study [27], the relationship between health status and lung function, as well as respiratory and peripheral muscle function, was examined. Correlations were found between respiratory muscle endurance time and the health status aspects of mobility and body care and movement. In contrast, others found also skeletal muscle weakness in patients with sarcoidosis who complained of fatigue. This weakness was related to reduced health status and exercise intolerance [29]. The radiographic stage was related to cognitive and emotional behavior, home management, and social interaction [27]. With regard to the relationship between pulmonary function tests and dyspnea, and health status, results are inconsistent. Yeager *et al.* [30] found that lower scores on the

spirometric tests and more self-reported dyspnea were related to a diminished health status. In another, smaller, study [28] those relationships were not found. Finally, Baughman *et al.* [31] examined the usefulness of fluticasone in patients with acute symptomatic pulmonary sarcoidosis. No difference was found between the fluticasone ($n=10$) and the placebo ($n=11$) group with regard to health status. Oral corticosteroids appeared to be associated with significant complaints, however, while inhaled corticosteroids were well tolerated [31].

Idiopathic pulmonary fibrosis

Concerning health status and IPF, a systematic review has been made by Swigris *et al.* [32]. From the seven studies they included in their review, two [11,23] mainly concerned QOL. These studies have been discussed previously.

Swigris *et al.* [32] found that, compared with patients with moderately severe chronic obstructive pulmonary disease, patients with moderately severe IPF had similar scores on the St George Respiratory Questionnaire. IPF patients experienced most problems with the activity domain (St George Respiratory Questionnaire). Although the review showed that patients' health status was worse in almost every domain in comparison with healthy persons, patients experienced the most impairment in the physical domain. Overall, the correlations between health status and pulmonary function tests were low [32]. Clark *et al.* [33], however, found that forced vital capacity was predicted by mastery, physical functioning, energy, health perceptions and fewer depressive symptoms. This study had a high nonresponse rate and a substantial number of patients also suffered from rheumatoid arthritis. A study [34] examining the relationship between health status and dyspnea scales found substantial to high correlations. Although the authors concluded that dyspnea scales can be used as health status measures, the common variance only exceeded 50% with regard to physical functioning. This percentage was much lower for associations between dyspnea and psychological and social functioning [34]. Others concluded that dyspnea scales and the St George Respiratory Questionnaire, and not the Quality of Well-being questionnaire, are useful tools for measuring health status in IPF patients; however, this conclusion was solely based on the significant correlations with pulmonary function tests and arterial blood gas measures at rest and during exercise [13]. In a study into nocturnal hypoxemia [33], physical and social functioning, and energy were related to abnormal sleep and lower overnight S_aO_2 . With regard to the effect of treatment on health status, no significant difference has been found between patients who received antioxidants or *N*-acetylcysteine [35]; interferon- γ also showed no effect on patients' health status [9,36]. A study [37]

focusing on factors contributing to patients' health status revealed that dyspnea was the only predictor.

Discussion

The aim of this review was to provide information on the influence of ILD on patients' health status and QOL, with an emphasis on which aspects might be affected by ILD.

In the literature, two distinct concepts are examined under the heading QOL: health status, often erroneously called health-related QOL, and QOL. While health status concerns the impact of disease on functioning, (health-related) QOL also reflects patients' evaluation of their functioning. It is important to make this distinction when planning and performing intervention studies, because health status and QOL measures may produce different results.

Almost all studies concern sarcoidosis or IPF. Results from the existing studies show that both illnesses have a substantial impact on patients' life. The QOL of sarcoidosis patients with current symptoms and patients with IPF is mainly impaired in the domains of physical health and level of independence. Concerning health status, sarcoidosis patients report reduced physical functioning, especially when they have current symptoms. The health status of IPF patients is impaired in almost all aspects. In sarcoidosis, the major problem is fatigue; in IPF, dyspnea, cough and exercise intolerance are devastating, and have a substantial impact on patients' lives. In IPF, subjectively assessed dyspnea is related to health status. A relationship between treatment and health status has not been found.

Studies aiming at measuring QOL or health status in other ILDs are scarce or nonexistent. Future studies are needed to obtain information on aspect of life in these patients that are affected by their disease.

Except for the treatment trials that assessed health status, studies are cross-sectional in nature. The major disadvantage of such type of research is that it provides no information on the course of QOL and health status across time, making statements about cause and effect impossible. Follow-up studies focusing on QOL and/or health status are needed.

Conclusion

We have reviewed the influence of ILD on patients' health status and QOL. There appeared to be relatively few studies examining the health status or QOL of patients with ILD. Except for two studies, existing studies all concerned sarcoidosis and IPF. Research has shown that the health status and QOL of sarcoidosis and IPF patients are impaired. Most of these studies have a cross-sectional design. Concerning other ILDs,

there is a paucity of research studying health status and QOL.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- of special interest
- of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 375).

- 1 Loddenkemper R, Kloppenborg A, Schoenfeld N, *et al.* Clinical findings in 715 patients with newly detected pulmonary sarcoidosis: results of a cooperative study in former West Germany and Switzerland. WATL Study Group Wissenschaftliche Arbeitsgemeinschaft für die Therapie von Lungenkrankheiten Sarcoidosis. *Vasc Diffuse Lung Dis* 1998; 15:178–182.
- 2 Klonoff EA, Kleinhenz ME. Psychological factors in sarcoidosis: the relationship between life stress and pulmonary function. *Sarcoidosis* 1993; 10: 118–124.
- 3 Hoitsma E, Marziniak M, Faber CG, *et al.* Small fiber neuropathy in sarcoidosis. *Lancet* 2002; 359:2085–2086.
- 4 Wirsberger RM, De Vries J, Wouters EF, Drent M. Clinical presentation of sarcoidosis in The Netherlands: an epidemiological study. *Neth J Med* 1998; 53:53–60.
- 5 Drent M, Wirsberger RM, Breteler MH, *et al.* Quality of life and depressive symptoms in patients suffering from sarcoidosis. *Sarcoidosis Vasc Diffuse Lung Dis* 1998; 15:59–66.
- 6 Wirsberger RM, De Vries J, Breteler MH, *et al.* Evaluation of quality of life in sarcoidosis patients. *Respir Med* 1998; 92:750–756.
- 7 De Vries J, Wirsberger RM. Fatigue, quality of life, and health status in sarcoidosis. *Eur Respir Monogr* 2005; 10:92–104.
- 8 Bitterman PB, Rennard SI, Keogh BA, *et al.* Familial idiopathic pulmonary fibrosis: evidence of lung inflammation in unaffected family members. *N Engl J Med* 1986; 314:1343–1347.
- 9 Abdelaziz MM, Samman YS, Wali SO, Hamad MM. Treatment of idiopathic pulmonary fibrosis: is there anything new? *Respirology* 2005; 10:284–289.
- 10 De Vries J, Seebregts A, Drent M. Assessing health status and quality of life in idiopathic pulmonary fibrosis: which measure should be used? *Respir Med* 2000; 94:273–278.
- 11 De Vries J, Kessels BLJ, Drent M. Quality of life of idiopathic pulmonary fibrosis patients. *Eur Respir J* 2001; 17:954–961.
- 12 Hope-Gill BDM, Hilldrup S, Davies C, *et al.* A study of the cough reflex in idiopathic pulmonary fibrosis. *Am J Respir Crit Care Med* 2003; 168:995–1002.
- 13 Tzanakis N, Samiou M, Lambiri I, *et al.* Evaluation of health-related quality of life and dyspnea scales in patients with idiopathic pulmonary fibrosis: correlation with pulmonary function tests. *Eur J Intern Med* 2005; 16:105–112.
- 14 Madison JM, Irwin RS. Chronic cough in adults with interstitial lung disease. *Curr Opin Pulm Med* 2005; 11:412–416.
- 15 De Vries J. Quality of life assessment. In: Vingerhoets AJJM, editor. *Assessment in behavioural medicine*. Hove: Brunner-Routledge; 2001. pp. 353–370.
- 16 Curtis JR, Patrick DL. The assessment of health status among patients with COPD. *Eur Respir J* 2003; 41:36s–45s.
- 17 WHOQOL Group. Development of the WHOQOL: rationale and current status. *Int J Ment Health* 1994; 23: 24–56.
- 18 WHO. The first 10 years of the World Health Organization. Geneva: WHO; 1958.
- 19 De Vries J, Drent M. Quality of life and health status in sarcoidosis. In: Baughman R, editor. *Sarcoidosis*. London: Taylor & Francis; 2006. pp. 463–475.
- 20 De Vries J, Drent M, Van Heck GL, Wouters EFM. Quality of life in sarcoidosis: a comparison between members of a patient organization and a random sample. *Sarcoidosis Vasc Diffuse Lung Dis* 1998; 15:183–188.
- 21 De Vries J, Van Heck GL, Drent M. Gender differences in sarcoidosis: symptoms, quality of life and medical consumption. *Women Health* 1999; 30:99–114.
- 22 Wirsberger RM, De Vries J, Jansen TLThA, *et al.* Impairment of quality of life: rheumatoid arthritis versus sarcoidosis. *Neth J Med* 1999; 54:86–95.
- 23 De Vries J, Seebregts A, Drent M. Assessing health status and quality of life in idiopathic pulmonary fibrosis: which measure should be used? *Respir Med* 2000; 94:273–278.

- 24 Reinhold-Keller E, Herlyn K, Wagner-Bastmeyer R, *et al.* Effect of Wegener's granulomatosis on work disability, need for medical care, and quality of life in patients younger than 40 years at diagnosis. *Arthritis Rheum* 2002; 47: 320–325.
  - 25 Tang WK, Lum CM, Ungvari GS, Chiu HF. Alcohol consumption, lung function, and quality of life in pneumoconiosis. *Alcohol Clin Exp Res* 2005; 29:1230–1236.
  - 26 Chang JA, Curtis JR, Patrick DL, Raghu G. Assessment of health-related quality of life in patients with interstitial lung disease. *Chest* 1999; 116: 1175–1182.
  - 27 Wirnsberger RM, Drent M, Hekelaar N, *et al.* Relationship between respiratory muscle function and quality of life in sarcoidosis. *Eur Respir J* 1997; 10: 1450–1455.
  - 28 Cox CE, Donohue JF, Brown CD, *et al.* Health-related quality of life of persons with sarcoidosis. *Chest* 2004; 125:997–1004.
  - 29 Spruit MA, Thomeer MJ, Gosselink R, *et al.* Skeletal muscle weakness in patients with sarcoidosis and its relationships with exercise intolerance and reduced health status. *Thorax* 2004; 60:32–38.
  - 30 Yeager H, Rossman MD, Baughman RP, *et al.*, ACCESS Research Group.
    - Pulmonary and psychosocial findings at enrollment in the ACCESS study. *Sarcoidosis Vasc Diffuse Lung Dis* 2005; 22:147–153.
 Lung involvement and the association of demographic and psychosocial factors with respiratory health were examined in 736 persons with sarcoidosis. Impaired spirometry and greater dyspnea were associated with poorer quality of life.
  - 31 Baughman RP, Iannuzzi MC, Lower EE, *et al.* Use of fluticasone in acute symptomatic pulmonary sarcoidosis. *Sarcoidosis Vasc Diffuse Lung Dis* 2002; 19:198–204.
  - 32 Swigris JJ, Kuschner WG, Jacobs SS, *et al.* Health-related quality of life in patients with idiopathic pulmonary fibrosis: a systematic review. *Thorax* 2005; 60:588–594.
- A systematic review to examine what is known about QOL and health status in patients with IPF. IPF affected physical health, general health, energy level, respiratory symptoms and level of independence.
- 33 Clark M, Cooper B, Singh S, *et al.* A survey of nocturnal hypoxaemia and health related quality of life in patients with cryptogenic fibrosing alveolitis. *Thorax* 2001; 56:482–486.
  - 34 Baddini Martinez JA, Martinez TY, Lovetro Galhardo FP, de Castro Pereira CA. Dyspnea scales as a measure of health-related quality of life in patients with idiopathic pulmonary fibrosis. *Med Sci Monit* 2002; 8:CR405–CR410.
  - 35 Tomioka H, Kuwata Y, Imanaka K, *et al.* A pilot study of aerosolized *N*-acetylcysteine for idiopathic pulmonary fibrosis. *Respirology* 2005; 10: 449–455.
  - 36 Raghu G, Brown KK, Bradford WZ, *et al.*, Idiopathic Pulmonary Fibrosis Study Group. A placebo-controlled trial of interferon gamma-1b in patients with idiopathic pulmonary fibrosis. *N Engl J Med* 2004; 350:125–133.
  - 37 Nishiyama O, Taniguchi H, Kondoh Y, *et al.* Health-related quality of life in patients with idiopathic pulmonary fibrosis: what is the main contributing factor? *Respir Med* 2005; 99:408–414.