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Managing Dyspnea in Individuals With Idiopathic Pulmonary Fibrosis

Kathryn E. Fenwick, BSN, RN ❍ Beth Ann Swan, PhD, RN, FAAN

Idiopathic pulmonary fibrosis is an unrelenting form of interstitial lung disease associated with a high symptom burden and reported low health-related quality of life. Clinicians have access to limited pharmacologic interventions to help slow the disease progression. Nonpharmacologic interventions are vital in managing dyspnea for these individuals, which is one of the most frequently reported factors that negatively impacts health-related quality of life. Common methods of symptom control include integration of pulmonary rehabilitation, supplemental oxygen, and interdisciplinary support, such as support groups, palliative care, and case conferences, into routine medical care. This literature review describes a multidisciplinary approach for managing dyspnea to improve health-related quality of life for those with idiopathic pulmonary fibrosis. Findings demonstrate that structured pulmonary rehabilitation programs, fast-track case conferences, and supplemental oxygen therapy are most effective. Further research is needed to demonstrate a clinically significant benefit of palliative care visits in the long term for these individuals.

KEY WORDS
dyspnea, health-related quality of life, idiopathic pulmonary fibrosis, interstitial lung disease, palliative care

The term *interstitial lung disease* encompasses a wide range of parenchymal lung conditions that affect the pulmonary interstitium characterized by inflammation and pulmonary fibrosis, or lung scarring. It is widely recognized that treatment for specific types of interstitial lung disease depends on the extent of lung damage, as it is irreversible and progressive.[1] In cases such as idiopathic pulmonary fibrosis, the most common and arguably the most aggressive form of pulmonary fibrosis, individuals are faced with a survival rate of a mere 2 to 3 years after diagnosis, high symptom burden, and reported poor quality of life.[2,3] Treatment methodologies often rely on slowing disease progression, relieving symptom burden, and improving quality of life.[1] Specialized health care networks and providers to assist individuals with idiopathic pulmonary fibrosis with proper or timely treatment are scarce.[4] Individuals are often misdiagnosed for an average of 1.5 years, worsening unmet needs regarding psychological support, treatment plans, and symptom management.[4] These concerns, unfortunately, are often prevalent even after an accurate diagnosis is made.

A lack of treatment options remains a concern when addressing how to improve health-related quality of life for these individuals. Individuals with idiopathic pulmonary fibrosis frequently report an unrelenting dry cough and progressive dyspnea with exertion, which severely encumbers their ability to be active participants in their daily lives, thus reducing health-related quality of life.[5] Beneficial components of treatment reported by individuals with idiopathic pulmonary fibrosis include high-quality education on disease progression, emotional support, timely and accurate diagnosis, supportive end-of-life care planning, collaboration with a knowledgeable health care team, and both pharmacologic and nonpharmacologic methods of symptom management.[6] The purpose of this literature review is to describe the multidimensional management of dyspnea in individuals with idiopathic pulmonary fibrosis.

SEARCH STRATEGY

The databases CINAHL, Cochrane Library Databases, PubMed, Scopus, and Google Scholar were queried between January 15 and January 21, 2020. The following search terms were used alone or in combination: "dyspnea," "breathlessness," "interstitial lung disease," "idiopathic pulmonary fibrosis," "quality of life," and "pulmonary fibrosis." In addition, manual searches of relevant research were carried out to review pertinent studies and provide a comprehensive search. Only peer-reviewed journal articles in English published within the last 5 years in academic journals that focused on idiopathic pulmonary fibrosis populations were included. Of the initial search, articles were omitted for the following reasons: individual involvement in lung transplantation, no standard measurement for health-related quality of life, and use of inpatient settings.

Kathryn E. Fenwick, BSN, RN, is post BSN to DNP student, Jefferson College of Nursing, Thomas Jefferson University, Philadelphia, PA.

Beth Ann Swan, PhD, RN, FAAN, is clinical professor and associate dean and vice president for academic practice partnerships, Nell Hodgson Woodruff School of Nursing, Emory University, Atlanta, GA.

Address correspondence to Kathryn E. Fenwick, 743 S 20th St, Apt 3, Philadelphia, PA 19146 (kef023@jefferson.edu).

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Thirteen articles are included in this literature review, including 7 randomized controlled trials and 6 observational studies of various designs.

SYNTHESIS OF LITERATURE

The 13 articles were sorted based on the intervention used to alleviate dyspnea and impact self-reported health-related quality of life as a primary or secondary outcome in individuals with idiopathic pulmonary fibrosis. The operational definition of quality of life for all studies used reliable and valid health-related quality of life assessments including (1) Saint George's Respiratory Questionnaire, (2) Saint George's Respiratory Questionnaire IPF-specific version, (3) King's Brief Interstitial Lung Disease Questionnaire, (4) Short Form 36 Health Survey, (5) University of California San Diego Shortness of Breath Questionnaire, (6) Visual Simplified Respiratory Questionnaire, (7) Psychological General Well-Being Index, and (8) Chronic Respiratory Disease Questionnaire.

Five articles assessed the impact of outpatient facility–based pulmonary rehabilitation or exercise programs on health-related quality of life assessments.[7-11] Two articles assessed the impact of outpatient home-based pulmonary rehabilitation or exercise programs on health-related quality of life assessments.[12,13] Unstructured exercise, or daily activity level, was evaluated by 2 groups.[14,15] The impact of supplemental oxygen was assessed in 1 article.[16] Three studies analyzed the effect of various methods of multidisciplinary support on pulmonary rehabilitation, including palliative care, fast-track case conferences, and support groups.[17-19] For organizational purposes of this review, the studies are grouped into 3 categories based on the overarching intervention used to impact health-related quality of life: (1) exercise to improve dyspnea, (2) intensive interdisciplinary support to improve dyspnea, and (3) supplemental oxygen to improve dyspnea (Table).

Managing Dyspnea With Exercise

Recommended supportive measures to help individuals manage dyspnea, and subsequently improve health-related quality of life, include pulmonary rehabilitation.[5] Pulmonary rehabilitation has been shown to assist individuals in gaining more control over their symptom burden of dyspnea and associated depression, anxiety, and even feelings of loss of control over one's life.[6] However, as it is viewed as a treatment option primarily for chronic obstructive pulmonary disease (COPD), there is often a disparity in pulmonary fibrosis individuals' ability to access pulmonary rehabilitation programs in their area.[20] The most updated American Thoracic Society guidelines for the treatment and management of idiopathic pulmonary fibrosis advise that pulmonary rehabilitation programs are not appropriate for all idiopathic pulmonary fibrosis patients.[3] Pulmonary rehabilitation programs may need to be explicitly tailored to the individual's idiopathic pulmonary fibrosis needs and take into consideration the degree of baseline functional status.[3] There are inconsistencies as to what length, regimen, or educational component of pulmonary rehabilitation programs is most beneficial when tailored for individuals with interstitial lung disease.

Severe exertional dyspnea in individuals with idiopathic pulmonary fibrosis can inhibit their desire to complete simple daily tasks, often noted as a key determinant of their health-related quality of life.[21] An improvement in idiopathic pulmonary fibrosis individually reported health-related quality of life has been found to have a robust link with improvement in clinical outcomes, including rates of lung function decline, hospitalization, and acute exacerbations of disease.[6]

Outpatient Facility–Based Pulmonary Rehabilitation

Outpatient facility–based pulmonary rehabilitation programs with short-term durations ranging from 8 to 12 weeks were the most commonly evaluated intervention to reduce dyspnea and improve health-related quality of life.[7-9,11] Arizono et al,[7] Dowman et al,[8] and Naz et al[9] assessed short-term effects of pulmonary rehabilitation using a validated questionnaire for self-reported quality of life, the Saint George's Respiratory Questionnaire, with Dowman et al[8] further specifying the questionnaire to be the idiopathic pulmonary fibrosis–specific version. Findings from these 3 studies demonstrated that short-term pulmonary rehabilitation programs produced significant improvement in health-related quality of life evidenced by improved parameters on the Saint George's Respiratory Questionnaire. Naz et al[9] provided evidence that short-term pulmonary rehabilitation can improve all subparameters of Saint George's Respiratory Questionnaire, whereas Dowman et al[8] demonstrated improvement in all subparameters except for Saint George's Respiratory Questionnaire-impact score. Arizono and colleagues[7] showed evidence of significantly improved Saint George's Respiratory Questionnaire total scores for individuals with idiopathic pulmonary fibrosis when compared with a control group of COPD patients. Interestingly, Naz and colleagues[9] further demonstrated that the significant improvement in Saint George's Respiratory Questionnaire scores was maintained and even more profound at 12 weeks than 8 weeks, demonstrating a continued positive effect.

Two randomized controlled trials evaluated the long-term effects of pulmonary rehabilitation on health-related quality of life using the Saint George's Respiratory Questionnaire.[10,11] Vainshelboim and colleagues[11] demonstrated that a 12-week supervised pulmonary rehabilitation program had a statistically significant improvement for both Saint George's Respiratory Questionnaire-total and Saint George's Respiratory Questionnaire-impact scores after 11 months. Perez-Bogerd and colleagues[10] used a 6-month pulmonary rehabilitation program design, resulting in clinically

significant improvement in Saint George's Respiratory Questionnaire activity scores, as well as Chronic Respiratory Disease Questionnaire dyspnea, emotion, and mastery scores. This clinically meaningful difference was present even at the 1-year follow-up assessment.[10] Thus, both studies, assessing a shorter-duration pulmonary rehabilitation program of 12 weeks and a longer-duration pulmonary rehabilitation program of 6 months, demonstrated maintenance of health-related quality of life outcomes for individuals with idiopathic pulmonary fibrosis in the long term. Interestingly, changes in 6-minute walk distance, a commonly monitored indicator of lung function in individuals with idiopathic pulmonary fibrosis, were associated with changes in reported health-related quality of life.[11] Although the studies reviewed supported facility-based pulmonary rehabilitation, home-based pulmonary rehabilitation is also recognized as an intervention for symptom management of dyspnea.

Outpatient Home-Based Pulmonary Rehabilitation

Two studies analyzed the effects of outpatient home-based pulmonary rehabilitation programs and its subsequent impact on health-related quality of life for individuals with idiopathic pulmonary fibrosis.[12,13] In an observational study with retrospective analysis, Wallaert and colleagues[12] demonstrated that a structured 2-month pulmonary rehabilitation program with once-weekly visits for education and psychosocial support had clinical improvement in self-reported health-related quality of life at 2-, 6-, and 12-month follow-up intervals using the Visual Simplified Respiratory Questionnaire. Of note, disease severity did not impact Visual Simplified Respiratory Questionnaire scores between groups.[12] In contrast, Yuen and colleagues[13] used a less structured exercise format of using a home-based exercise game involving full-body movements 3 times weekly. After 12 weeks, this regimen failed to yield any clinically significant improvement in health-related quality of life, as demonstrated by self-reported Saint George's Respiratory Questionnaire scores.[13] In addition to structured facilitated and home-based pulmonary rehabilitation, unstructured activity has also been evaluated in relation to the symptom management of dyspnea in individuals with idiopathic pulmonary fibrosis.

Unstructured Activity

Nishiyama et al[14] used a prospective controlled observational study to evaluate the impact of physical activity on individuals with idiopathic pulmonary fibrosis in an outpatient setting; idiopathic pulmonary fibrosis patients used an activity monitor for 7 days, with assessments of health-related quality of life through Saint George's Respiratory Questionnaire reporting at baseline and completion. There was no clinically significant correlation between physical activity variables, including steps walked and activity energy expenditure on Saint George's Respiratory Questionnaire scores.[14] Similarly, in a prospective cohort study, Root et al[15] evaluated the association between physical activity and health-related quality of life in individuals with pulmonary fibrosis of varying etiology who did not use supplemental oxygen by tracking activity for 7 consecutive days. No causation was established between physical activity, measured in steps per day, and improved health-related quality of life scores, as assessed with a variety of health-related quality of life tools, namely, University of California San Diego Shortness of Breath Questionnaire and Short Form 36 Health Survey.[15] Both studies provide support that physical activity alone, such as exercise that is not performed in a formal pulmonary rehabilitation setting, is not enough to have clinically meaningful impacts on self-reported health-related quality of life for individuals with idiopathic pulmonary fibrosis.[14,15] However, it adds to the discussion of what aspects of pulmonary rehabilitation programs may influence clinically significant improvements in quality of life that general physical activity does not. Whereas pulmonary rehabilitation is an important part of managing idiopathic pulmonary fibrosis, interdisciplinary care teams are another strategy for supporting symptom management of dyspnea in individuals with idiopathic pulmonary fibrosis.

Improving Dyspnea Symptom Management With Interdisciplinary Support

The most effective interventions to help manage symptom burden, particularly that of dyspnea, are multidisciplinary in nature and encompass both physical and psychosocial needs.[5] These approaches are accomplished most effectively through multidisciplinary coordination of care to provide individuals with treatment options.[22] Specialized interstitial lung disease registered nurses are crucial resources for individuals with idiopathic pulmonary fibrosis during discussions of palliative care, symptom burden, oxygen use, and methods of increasing abilities to participate in activities of daily living.[20] Early palliative care referrals are noted as a recommended course of action to help individuals with idiopathic pulmonary fibrosis understand the disease process and associated symptom burden.[23-25]

The use of various methods of providing more intensive direct multidisciplinary support to help individuals better understand and cope with breathlessness was assessed by 3 studies.[17-19] Janssen and colleagues[18] assessed the impact of structured palliative care visits at 3 and 6 months integrated into standard of care clinic visits in a randomized controlled trial. Magnani and colleagues[19] analyzed the impact of attending an idiopathic pulmonary fibrosis-specific support group for 6 months in a quasi-experimental pretest-posttest study. Bajwah and colleagues[17] evaluated the use of implementing a fast-track case conference, Hospital2Home, in a randomized controlled trial.

TABLE Summary of Findings

Method of Dyspnea Management		Citation	Population	Design/Methods	HRQOL Measurement Tool	Relevant Results and Findings
Exercise	Outpatient facility–based PR	Arizono et al,[7] 2017	N = 47 • 22 IPF • 27 COPD • Outpatient clinic (Japan)	• Prospective observational study • The IPF group (n = 24) and COPD group (n = 28) completed a 10-wk PR program. • Evaluations at baseline and 10 wk	SGRQ	• SGRQ-total scores with a statistically significant improvement for IPF and COPD patients after 10 wk of PR • No clinically significant difference between the IPF and COPD groups
		Dowman et al,[8] 2017	N = 142 • 61 IPF • 22 asbestosis • 23 connective tissue disease–related ILD • 36 other ILD etiologies • Three outpatient clinics (Australia)	• Randomized controlled trial • The intervention group (n = 74) completed an 8-wk PR program. • The control group (n = 68) received routine care. • Evaluations at baseline, 9 wk, and 6 mo	CRQ SGRQ-I	• The PR group demonstrated clinically significant short-term improvements in all CRDQ and SGRQ-I domains, except SGRQ-I-impact score. • PR is effective in patients across a range of ILDs with clinically meaningful benefits in asbestosis and IPF.
		Naz et al,[9] 2018	N = 28 • 14 ILD • 11 IPF • 2 sarcoidosis • 1 NSIP • PR unit associated with ILD clinic	• Prospective uncontrolled study • All participants completed a 12-wk PR program. • Evaluations at baseline, 8 wk, and 12 wk	SGRQ SF-36	• All SGRQ domains showed a significant improvement at weeks 8 and 12. • At week 12 of PR, patients had improved HRQOL compared with week 8 evaluations. • Significant improvement in mental health and vitality domains of SF-36 at week 8
		Perez-Bogerd et al,[10] 2018	N = 60 • IPF only • Specialty clinic (Belgium)	• Randomized controlled trial • The intervention group (n = 30) completed a 6-mo PR program. • The control group (n = 30) received routine care. • Evaluations at baseline, 3 mo, 6 mo, and 12 mo	SGRQ CRQ	• Clinically significant improvement shown after 3 mo of PR for SGRQ-activity domain and CRQ-dyspnea, CRQ-emotion, and CRQ-mastery scores • These improvements in HRQOL exceeded the minimal clinically important difference after PR and at 1-year mark.
		Vainshelboim et al,[11] 2015	N = 34 • IPF only • Outpatient clinic	• Randomized controlled trial • The intervention group (n = 16) completed a 12-wk supervised PR program in addition to routine care. • The control group (n = 18) received routine care. • Evaluations at baseline and 11 mo	SGRQ	• Statistically significant improvement for SGRQ-total and SGRQ-impact scores after 11 mo, demonstrating maintenance of outcomes in the long term. • Changes in HRQOL associated with improved walking capacity

(continues)

TABLE Summary of Findings, Continued

Method of Dyspnea Management		Citation	Population	Design/Methods	HRQOL Measurement Tool	Relevant Results and Findings
	Outpatient home-based PR	Wallaert et al,[12] 2019	N = 112 • 61 IPF • 51fibrotic NSIP • Specialty outpatient center	• Observational study with retrospective analysis • All completed a 2-mo PR program in home with visit for education/psychosocial support each week. • Evaluations before, upon completion, 6 mo, and 12 mo	VSRQ	• VSRQ scores showed a clinically significant improvement after a 2-mo program and evaluations at the 6- and 12-mo mark. • No effect of disease severity (mild to moderate vs severe) on VSRQ scores
		Yuen et al,[13] 2019	N = 20 • IPF only • Specialty clinic (UAE)	• Randomized, single-blinded controlled trial • The intervention group (n = 10) completed home-based Wii game with whole-body movements at least 3 times weekly. • The control group (n = 10) was assigned to Wii game with no whole-body movements. • Evaluations at baseline and 12-wk mark	SGRQ	• No clinically significant improvement in SGRQ scores between the intervention and control groups
	Unstructured activity	Nishiyama et al,[14] 2018	N = 51 • 31 IPF • 20 age-matched healthy • Outpatient clinic (Japan)	• Prospective controlled observational study • All participants used activity monitor for 7 consecutive days • Evaluations at baseline and upon completion	SGRQ	• No significant correlation between physical activity variables (steps walked or activity energy expenditure) and SGRQ scores
		Root et al,[15] 2017	N = 194 • Pulmonary fibrosis of any etiology • ILD clinic or online recruitment	• Prospective cohort study • All completed questionnaires at enrollment • All used accelerometer/GPS tracker for 7 d	UCSDSOBQ SF-36	• Causation not established between activity and HRQOL • Better daily physical activity (steps walked) associated with better HRQOL scores

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TABLE Summary of Findings, Continued

Method of Dyspnea Management	Citation	Population	Design/Methods	HRQOL Measurement Tool	Relevant Results and Findings
Interdisciplinary support	Bajwah et al,[17] 2015	N = 53 • Advanced idiopathic fibrotic lung disease only • Specialist outpatient clinic (London)	• Randomized controlled trial with qualitative interviews • The intervention group (n = 26) received fast-track home case conference (Hospital2Home) alongside routine care • The control group (n = 27) with routine care, placed onto 4-wk waitlist for case conference • Data collected at baseline, week 4, and week 8	KBILD SGRQ	• Statistical improvement in HRQOL after 4 wk, with effects being maintained at 8 wk • SGRQ-impact and SGRQ-total scores improved for the intervention group. • Positive effect on patients and caregivers of Hospital2Home intervention on HRQOL • SGRQ scores showed a similar effect as the intervention group when the control group received intervention.
	Janssen et al,[18] 2020	N = 22 • IPF only • Outpatient specialty clinic	• Randomized controlled trial • The intervention group (n = 11) received routine care with structured palliative care visits at 3 and 6 mo. • The control group (n = 11) received routine care alone. • Evaluations at baseline, 3 mo, and 6 mo	SGRQ	• Receiving palliative care may worsen HRQOL in the short term. • May cause worsening transient depression at 3 mo, but not at 6 mo
	Magnani et al,[19] 2017	N = 18 • IPF only • Specialty clinic (Italy)	• Quasi-experimental pretest-posttest study • All individuals participated in an IPF-specific support group conducted by nurses. • Evaluations at baseline and 6 mo of attendance	PGWBI	• Null association between support group attendance and psychosocial well-being
Supplemental oxygen	Visca et al,[16] 2018	N = 84 • Fibrotic interstitial lung disease only • Three specialty outpatient centers (UK)	• Prospective, open-label, mixed-method, crossover randomized control trial • The intervention group (n = 41) received oxygen first. • The control group (n = 43) received no oxygen at the start. • Evaluations at baseline, 2 wk, and 4 wk • Qualitative interviews completed at the end of trial	KBILD SGRQ UCSDSOBQ	• Initial use of ambulatory oxygen demonstrated a clinically significant improvement in KBILD-total, KBILD-breathlessness, KBILD-chest symptoms, UCSDSOBQ, SGRQ-total, and SGRQ-activity scores. • No clinically significant improvement associated between ambulatory oxygen and KBILD-psychological symptom score

Abbreviations: COPD, chronic obstructive pulmonary disease; CRQ, Chronic Respiratory Disease Questionnaire; HRQOL, health-related quality of life; ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; KBILD, King's Brief Interstitial Lung Disease Questionnaire; NSIP, nonspecific interstitial pneumonia; PGWBI, Psychological General Well-Being Index; PR, pulmonary rehabilitation; SF-36, Short Form 36 Health Survey; SGRQ-I, Saint George's Respiratory Questionnaire IPF-specific version; SGRQ, Saint George's Respiratory Questionnaire; UAE, United Arab Emirates; UCSDSOBQ, University of California San Diego Shortness of Breath Questionnaire; UK, United Kingdom; VSRQ, Visual Simplified Respiratory Questionnaire.

There was no clinically beneficial impact of structured palliative care visits on health-related quality of life for individuals with mild idiopathic pulmonary fibrosis. However, the data do offer novel insights; palliative care may be helpful in enhancing health-related quality of life for those with more severe disease progression who are aware of their long-term outcome.[18] After 6 months of attendance at an idiopathic pulmonary fibrosis–specific support group, there was a null association between participation and psychosocial well-being and health-related quality of life, as demonstrated by the Psychological General Well-Being Index scores.[19] This study provided data that showed interesting increases in psychosocial well-being parameters that deserve further investigation in the use of support groups for enhancing health-related quality of life in larger sample sizes.[19] Hospital2Home implementation improved health-related quality of life after 4 and 8 weeks as evidenced by Saint George's Respiratory Questionnaire-impact and Saint George's Respiratory Questionnaire-total scores.[17] Similar improvements were seen when the control group received the Hosptial2Home addition to their routine medical care.[17] This demonstrated that early integration of home-based case conferences has a more valuable impact on health-related quality of life for idiopathic pulmonary fibrosis patients than either participation in idiopathic pulmonary fibrosis–specific support groups or early incorporation of palliative care visits.[17] Although interdisciplinary approaches show some promise for managing dyspnea, less is known about the use of oxygen.

Controlling Dyspnea With Supplemental Oxygen

Limited data exist to support the use of oxygen for palliative benefit, although it is recommended for severe breathlessness.[26,27] Visca et al[16] evaluated the short-term effects of using supplemental oxygen on health-related quality of life for individuals with fibrotic interstitial lung disease with exertional hypoxia through a prospective open-laboratory, mixed-method, crossover randomized controlled trial. Saint George's Respiratory Questionnaire and King's Brief Interstitial Lung Disease Questionnaire scores were evaluated at baseline and at 2- and 4-week follow-up points.[16] Supplemental oxygen was associated with clinically significant short-term improvement in the following subdomains: King's Brief Interstitial Lung Disease Questionnaire total, King's Brief Interstitial Lung Disease Questionnaire-breathlessness, King's Brief Interstitial Lung Disease Questionnaire-chest symptoms, Saint George's Respiratory Questionnaire total, and Saint George's Respiratory Questionnaire-activity.[16] Notably, there was no clinically significant improvement associated between supplemental oxygen use and King's Brief Interstitial Lung Disease Questionnaire-psychological symptom scores.[16] Three centers were used to recruit for this study, allowing for a diversity of study population. However, no other study used supplemental oxygen as an intervention. Despite negative perspectives on oxygen use, as evidenced by qualitative interviews and a low adherence rate of oxygen use during the study, early obtainment of supplemental oxygen for individuals' idiopathic pulmonary fibrosis with exertional hypoxia is an effective method of improving patient-reported health-related quality of life.[16]

GAPS IN LITERATURE AND LIMITATIONS

Limitations of this literature review include the use of studies with small sample sizes, some mixed interstitial lung disease populations, and subjective measures of reporting health-related quality of life. Larger randomized controlled trials should be used in future investigations to improve representation. Moreover, participants had varying use of antifibrotics, steroid medications, and supplemental oxygen. These were not always controlled for in all studies, limiting the applicability of these literature review findings to all individuals with idiopathic pulmonary fibrosis, such as those with mild, moderate, and severe diseases. Moreover, not all questionnaires used are validated specifically for idiopathic pulmonary fibrosis. For example, the Visual Simplified Respiratory Questionnaire used by Wallaert and colleagues,[12] is validated for use with persons with COPD but was used with an idiopathic pulmonary fibrosis sample because of ease of use and success in trials assessing home-based pulmonary rehabilitation in COPD patients. The studies reviewed provided interventions in the context of team-based care and did not focus on registered nurse–specific care interventions.

DISCUSSION

Emerging clinical research trials are looking at more innovative and effective methods of combatting and, hopefully, in the future, curing idiopathic pulmonary fibrosis. The most updated American Thoracic Society guidelines for treatment and management of idiopathic pulmonary fibrosis advise on the use of both pharmacologic and nonpharmacologic strategies.[3] There is no cure and only limited pharmacologic recommendations for treatment options. Therefore, nonpharmacologic interventions are key in providing comprehensive care for symptom management of idiopathic pulmonary fibrosis patients, particularly when managing dyspnea. The findings of this literature review support the American Thoracic Society recommendations but also help discern which nonpharmacologic interventions for dyspnea are more effective in improving health-related quality of life for individuals with idiopathic pulmonary fibrosis. Registered nurses can play an essential role in providing symptom management, palliative care, and behavioral health for these individuals.

Pulmonary rehabilitation programs are not recommended for all idiopathic pulmonary fibrosis patients.[3] Furthermore, evidence from this literature review supports that all pulmonary rehabilitation programs are not adequately designed to elicit effective responses for all individuals with idiopathic pulmonary fibrosis; outpatient facility–based and structured home-based pulmonary rehabilitation programs of any length were the most effective in improving health-related quality of life, with both clinically significant short- and long-term effects.[7-12] Activity alone in an unstructured and informal setting, such as at home, is not enough to provide clinically significant impacts on health-related quality of life.[13-15] Additional nonpharmacologic interventions to help improve dyspnea and, subsequently, improve health-related quality of life for individuals with idiopathic pulmonary fibrosis may include earlier use of supplemental oxygen for exertional dyspnea and fast-track case conferences in conjunction with their routine medical care.[16,17] Registered nurses should ensure that these nonpharmacologic interventions to alleviate dyspnea are included in routine management for persons with idiopathic pulmonary fibrosis to enhance health-related quality of life and improve clinical outcomes.

RECOMMENDATIONS FOR FUTURE RESEARCH

Future research opportunities exist in exploring both pulmonary rehabilitation program components and the impact of palliative care visits in the long term for individuals with idiopathic pulmonary fibrosis to improve health-related quality of life. To determine how to tailor pulmonary rehabilitation programs for idiopathic pulmonary fibrosis, researchers should explore the noted differences in pulmonary rehabilitation programs across facilities. This includes the length of sessions, frequency of sessions, monthly duration of programs, included educational components, and degree of pulmonary impairment before individuals are enrolled; this would aid in explaining why pulmonary rehabilitation programs, not physical activity alone, are beneficial in improving health-related quality of life. Studies with palliative care interventions were limited in both idiopathic pulmonary fibrosis disease progression (only focusing on individuals with mild disease) and time length (only short-term). Positive benefits have been shown in studies looking at the impact of early palliative care conversations focused on prognosis, symptom management, and advance care planning in the diagnosis of serious chronic illnesses in other populations.[28-30]

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