IMPROVING END-OF-LIFE CARE FOR PATIENTS WITH IDIOPATHIC PULMONARY FIBROSIS AND THEIR CARE PARTNERS

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Palliative care is increasingly recognized as relevant to the care of advanced disease in a variety of settings. Idiopathic pulmonary fibrosis (IPF) results in scarring of the lung, respiratory failure and, commonly, death within 3-5 years of diagnosis. The purpose of this study was to evaluate the impact of a 6-week program designed using palliative care concepts (PRISIM) on symptom burden and health-related quality of life (HRQoL) in patients with IPF and their care partners. Subjects were 42 participants randomized to an experimental (10 patients/care partners) or control (11 patients/care partners) group. The experimental group attended the 6-week PRISIM program and the control group received usual care. Prior to and immediately after attending the program, all participants completed questionnaires designed to assess anxiety, depression, perceived stress, and HRQoL. Participation in PRISIM decreased perceptions of physical HRQoL and tended to increase anxiety. Nevertheless, post course evaluations were highly positive. Post study qualitative interviews with experimental group participants yielded three common themes that reached saturation: “patients did not feel isolated”, were “able to put their disease into perspective”, and felt it “important to participate in research and help others”. Palliative interventions may increase anxiety but appear to yield other positive effects. Further exploration of the impact of such interventions is needed using both qualitative and quantitative methodology.

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PREFACE

There are many people who I would like to acknowledge as I complete this course of work, which has affectionately been coined a labor love. First I would like to thank my family. My husband and daughters have been extremely supportive as I toiled through this process. Next, my work family has provided me with a wealth of knowledge, as well as unending support to carry out this work. I would like to thank my advisor Leslie Hoffman who has guided me for many years and is just simply amazing. To be one of "Leslie's students" is an honor. I would also like to thank the people on my committee as they each helped me in their own unique way with their vast expertise. The patients and their families are the driving force for what I do. As I thought of this research, the underlying drive was to help them cope with the impact of this disease, Idiopathic Pulmonary Fibrosis.

I would like to dedicate this research to all nurses caring for patients with life threatening diseases in the hope that as they care for and cultivate all aspects of care required by the patient and their surrounding support systems, that they elevate the profession of nursing and provide comfort and support to the populations they serve.

1.0 INTRODUCTION

Interstitial lung disease (ILD) is a condition that results in progressive inability to maintain normal blood oxygen levels due to impaired transfer of gas across the alveolar-capillary membrane [1]. ILD includes a variety of conditions all of which share the common characteristics of lung scarring and progressive loss of the normal gas transfer ability [2]. Patients experience a progressive loss of functional ability and, ultimately, die from acute respiratory failure. More than 150 clinical diagnostic entities are associated with ILD [3] .

Idiopathic Pulmonary Fibrosis (IPF), also known pathologically as usual interstitial pneumonia (UIP), is one of the most common forms of ILD with an estimated 10.7 cases per 100,000 diagnosed each year for males and 7.4 cases per 100,000 for females [4]. In the absence of a surgical lung biopsy, the diagnosis of IPF remains uncertain. However, in the immunocompetent adult, the presence of all of the following major diagnostic criteria as well as at least three of the four minor criteria increases the likelihood of a correct clinical diagnosis of IPF. Major diagnostic criteria for IPF include: exclusion of other known causes of ILD, such as certain drug toxicities, environmental exposures, and connective tissue diseases; abnormal pulmonary function studies that include evidence of restriction (reduced VC often with an increased FEV1/FVC ratio) and impaired gas exchange [increased AaPO2 with rest or exercise or decreased DLCO]; bibasilar reticular abnormalities with minimal ground glass opacities on HRCT scans and transbronchial lung biopsy or bronchoalveolar lavage (BAL) showing no

features to support an alternative diagnosis. Minor criteria for diagnosis include: age > 50 yr; insidious onset of otherwise unexplained dyspnea on exertion; duration of illness ≥ 3 mo; and bibasilar, inspiratory crackles (dry or "Velcro" type in quality) [5].

IPF is often thought of as a disease with uniformly poor survival, but survival varies widely [6] [7]. Some patients die within one year of diagnosis, whereas others live longer than six years [8, 9]. The reason for this difference is unknown[9]. Although medications, including interferon gamma 1b, are being tested as a potential means of decreasing disability from IPF, there is currently no therapy that consistently reverses or cures the lung damage [10].

All patients diagnosed with ILD share problems common to those with a chronic illness. Chronic illness, a permanently altered health state, results from a non-reversible pathological condition [11]. The consequence is a residual disability that cannot be corrected by a simple surgical procedure or cured by a short course of medical therapy [12]. The diagnosis of a chronic illness can affect the emotional response, sometimes in a manner disproportionate to the extent of physical disability. This can be a major life change for the patient and family [13]. Consequently, it is essential to treat the psychosocial symptoms as well as the physical symptoms.

As with other chronic illnesses, the progressive disability associated with IPF impacts the patient and their care partner. The impact can be particularly devastating given the rapidly progressive nature of the decline, variability across individuals, and lack of effective therapy. DeVries and colleagues [14], in a study conducted in the Netherlands, assessed perceptions of health-related quality of life (HRQoL) in 41 patients with IPF and the relationship between QoL, depressive symptoms, and breathlessness in these patients. They reported that the QoL of patients with IPF was mainly impaired in the domains of “physical health” and “level of

independence" compared to a matched healthy controls. Of note, approximately 25% of patients with IPF indicated that they experienced significant depressive symptoms.

These findings suggest that patients with IPF may benefit from an advance care plan that provides assistance in coping with this decline[15]. Advance care planning is the process of initiating discussion, reflection, and understanding of one's health state in terms of goals, values, and preferences for future treatment [16]. The American Nurses Association (1991) states that nurses have a responsibility to "facilitate informed decision-making" that includes, but is not limited to, advanced directives [16]. The process of advance care planning begins with the patient's understanding of the disease and discussion about possible scenarios leading to death which, in itself may prepare the patient for the course of the disease [16].

The goals of treatment of IPF include managing symptoms and relieving discomfort. Palliative care should be offered from the time of diagnosis, even while the patient pursues treatments to slow or stabilize the disease [7]. Palliative care, also known as end-of-life care, refers to medical or nursing interventions that alleviate symptoms without improving or preventing progression of the underlying disease [17]. Dame Cicely Saunders, the Chair of St. Christopher's Hospice in London, the first inpatient, home care, research, and teaching hospice, is credited for her contributions to palliative care [18]. She states that palliative care "stems from the recognition of the potential at the end of life for discovering and for giving, a recognition that an important dimension of being human is the lasting dignity and growth that can continue through weakness and loss. The goal of palliative care is "to prevent and relieve suffering and to support the best possible quality of life for patients and their families, regardless of the stage of the disease or the need for other therapies"[19]. No member of the interdisciplinary team is more central to making these discoveries possible than the nurse" [18]. Although palliative care is

commonly integrated into the management of patients with cancer, its use in patients with advanced lung disease is more limited. Nevertheless, palliation is the only therapeutic option for many diseases of the lung, including lung cancer, chronic obstructive pulmonary disease, cystic fibrosis and IPF [17]. The concepts integral to palliative care seem particularly appropriate for patients with IPF who face a mortality rate that is greater than many cancers. The mantra "It is wise to hope for and expect the best, but it is also wise to prepare for the worst" is a way to introduce advance care planning to patients with IPF and their care partners [17].

No studies were identified that tested the ability of a brief intervention to assist patients with IPF to deal more effectively with their illness. Accordingly, we developed a 6-session *disease management intervention* (PRISIM: Program to Reduce Symptoms and Improve Lifestyle Management) designed to be delivered to the patient and care partner. This PRISIM program incorporated content taught to these patients by the Clinical Nurse Specialist, a program successfully used by Puskar [20] to assist a rural school age population to cope with depressive symptoms, and issues related to end-of-life planning. The PRISIM intervention was delivered over a six week period by a team of clinicians that included a pulmonary clinical nurse specialist, psychiatric clinical nurse specialist with training in cognitive behavior therapy, and a nurse certified as an advance care planning facilitator and instruction. Because chronic illness impacts the patient and care partner, both individuals were included as participants in all sessions of the program.

1.1 PURPOSE

The purpose of this study was to test the ability of a 6-week *disease management intervention* (PRISIM: Program to Reduce Symptoms and Improve Lifestyle Management) to decrease symptom burden, decrease perceived stress, and improve perceptions of health-related quality of life (HRQoL) for patients with IPF and their care partners. The proposed study employed a quantitatively driven, concurrent nested mixed-method design.

1.2 SPECIFIC AIMS

The specific aims of the study were to:

1) Describe the prevalence of anxiety and depression in patients with IPF and their care partners;

2) Determine the short term (6-week) impact of a disease management intervention on symptom burden (shortness of breath, anxiety, depression), perceived stress, and HRQoL in patients with IPF; and

3) Determine if participation in a disease management intervention altered caregiver burden as evidenced by changes in anxiety and depression, perceived stress, and HRQoL.

4) Discover the interpretive process of participation in the PRISIM program for patients and their care partners.

5) Evaluate the common themes of the experience of living with IPF from the perspective of the patient and care partner.

Hypothesis:

H1. Patients with IPF who participate in a disease management course will experience less anxiety, depression, perceived stress, and improved HRQoL.

H1. Care partners of patients with IPF who participate in a disease management course will experience less anxiety, depression, perceived stress, and improved HRQoL.

1.3 DEFINITION OF TERMS

1. Disease Management Intervention – intervention designed to assist patients to live life as fully as possible by reducing symptom burden, maintaining functional ability, and thereby achieving more optimal HRQoL.

2. Symptom Burden – perception of shortness of breath, anxiety, depression in the IPF patients; perception of anxiety and depression in the care partner.

3. Shortness of breath – scores on the *University of California at San Diego Shortness of Breath Questionnaire (SOBQ*, an index of the severity of shortness of breath during activities of daily living (ADL's) associated with varying levels of exertion.[21].

4. Anxiety – scores on the *Beck Anxiety Inventory (BAI)*, a standardized tool designed to assess the presence and severity of common symptoms of anxiety, both psychological and physical. [22].

5. Depression – scores on the *Beck Depression Inventory (BDI-II®),* a self-report instrument, is designed to measure the severity of depression in adults and adolescents 13 years or older [23].

6. Perceived stress – scores on the *Perceived Stress Scale,* a tool designed to measure the degree of perceived stress of a situation, and the degree to which subjects find their lives unpredictable, uncontrollable, and overloading. [24].

7. HRQoL – scores on the *SF-36 Short Form Medical Outcomes Study Form Version 2,* a widely used generic scale, that has been used to assess HRQoL in a variety of chronic medical conditions and validated in patients with IPF [25].

8. Idiopathic Pulmonary Fibrosis - also known pathologically as usual interstitial pneumonia (UIP) is one of the most common forms of ILD.

9. IPF Patient – patient diagnosed with IPF either radiologically or histologically by open lung biopsy.

10. Care Partner – patient's significant other (spouse, sibling, close friend) who is actively involved in care of patient with IPF.

1.4 SIGNIFICANCE TO NURSING

Patients with IPF and their care partners require advance care planning as they deal with the impact of the disease. As advance practice nurses, clinical nurse specialists have the capacity to serve an important role in the care of these individuals. An IPF Patient Support Group led by the Clinical Nurse Specialist in the Simmons Center for Interstitial Lung Disease at the University of Pittsburgh (KOL) fostered the belief that patients and their care partners were searching for more information about this disease than was currently being provided. The support group included educational sessions on a variety of topics including What is IPF?, Genetics of IPF, Treatment of IPF, Importance of Exercise and Pulmonary Rehabilitation, Home Oxygen Therapy, Anxiety,

Importance of Good Nutrition and was open to all patients seen at this Center. Discussions regarding other topics led to interest in developing an educational program that included information about coping with depressive symptoms and end-of-life planning. This conceptualization was innovative as educational programs for patients with advanced lung disease rarely include specific information on these topics, despite the high mortality rates of individuals with these conditions.

As a result of patient and care partner comments and course evaluations, a mixed methods approach was suggested to gather further information from the patients and care partners regarding how content presented in the course impacted their lives. We were particularly interested in exploring the experiences from the perspective of the patients and their care partners because, while they verbalized satisfaction with the experience, item scores suggested increased anxiety, compared to control group participants. A qualitative study was designed that incorporated home visits to experimental group participants 3-9 months after study completion to obtain qualitative data. Interviews of the patient and care partner were recorded, transcribed, and evaluated for common themes.

Information gained from this research is significant to nursing for several reasons. Increased awareness of the disease and the impact that it has on patients and their care partners is important for nurses who care for patients with IPF. Because this disease has a poor prognosis and no cure, nurses can help patients deal with the uncertainties and psychosocial aspects of the disease. The ability to enable people to find relief, support, and meaning at the end of their lives is at the core of nursing [18].

In an attempt to further clarify the cause and gain meaning from living with this disease, the warm autopsy program was initiated as a direct result of patient feedback from the support

group to the clinical nurse specialist.[26] This program serves to allow patients to donate their lungs at the end of their life for research purposes and was originated by a patient who “didn’t want others to suffer the way that he did”.

Nurses, in the role of advanced practice, such as clinical nurse specialist can coordinate support groups and intervention programs such as the PRISIM program that serve as an important source of care for the patient with IPF and their care partner(s) as they deal with the impact of the disease. Barriers such as distance, supplemental oxygen therapy, and lack of social support may affect the ability of many patients to attend weekly sessions in the PRISIM program, so a digital video disk (DVD) recording of the intervention is a possible means of improving the outreach and connecting with patients who are unable to attend. In addition, those who participate in these sessions can be given a DVD to review at home to reinforce concepts as need arises.

Patients with other types of advanced lung disease, e.g., end-stage chronic obstructive pulmonary disease, cystic fibrosis, lung cancer and pulmonary hypertension might additionally benefit from such an intervention as they also experience a progressive decline in functional ability and limited therapeutic options [8].

2.0 BACKGROUND AND SIGNIFICANCE

2.1 IDIOPATHIC PULMONARY FIBROSIS

Idiopathic pulmonary fibrosis (IPF) is a condition that results in scarring of the lung (fibrosis) and progressive inability to maintain normal blood oxygen levels due to impaired transfer of gas across the alveolar-capillary membrane [10], [27]. Current theories postulate that IPF results from repeated episodes of acute lung injury that, for unknown reasons, produce activation of alveolar epithelial cells, inflammation, aberrant healing of the injured area, and the development of pulmonary fibrosis [10]. Once present, the fibrosis is progressive with a consequent deterioration in exercise capacity, increasing dyspnea and hypoxemia and, ultimately, death from respiratory failure or a complicating condition, e.g., cardiac disease, lung cancer, or infection [10], [27]. The disease typically presents in the 6^{th} and 7^{th} decade of life [27]. Median survival after presentation is less than 3 years [10], [27]. IPF causes restrictive lung disease, i.e., a reduction in the forced vital capacity (FVC) and forced expiratory volume in one second (FEV_1) with a normal FEV_1/FVC. Using the FVC, severity of disease can be graded as mild (FVC > 70-80% pred), moderate (FVC 55-70% pred) or severe (FVC < 55% pred). With the exception of lung transplantation, there are no proven therapies for IPF. Unfortunately, most patients are not eligible for transplantation because of older age or complicating medical conditions. Of those eligible, one third die awaiting transplant [10].

2.2 ANXIETY AND DEPRESSION

Depression, one of the most prevalent and debilitating mental health conditions, affects 17.6 million Americans of all ages each year [28]. Comparisons of findings from surveys suggest there has been a 3-fold increase in population-wide depression in the United States between 1987 and 1997. However, it is unclear if this change represents an increase in the prevalence of depression or better diagnosis and treatment in the primary care setting. [29]. Regardless, it is well established that depression can increase morbidity and mortality in chronic illness [30]. Depression often coexists with other long-term health problems and complicates their management [28]. Depressive disease occurs across a continuum from mild, transient feelings of sadness to the severe, pervasive sense of helplessness and hopelessness that is characteristic of major depressive disorder (MDD) [31] . It is one of the most common psychiatric disorders in the United States, and although episodes may be of long duration, with a 50% rate of recurrence, depression is highly treatable [31]. The effects can be detrimental on quality of life and daily function and match those of heart disease, and exceed those of diabetes, arthritis, and peptic ulcer disease [32]. About 60 percent of depressed individuals have at least one other chronic medical condition, such as a heart problem, high blood pressure, or diabetes [29].

Depressive symptoms are commonly reported by patients with potentially terminal conditions. In 987 patients with lung cancer, the overall prevalence of self-rated depression was 33%, with a higher prevalence in patients with a poorer prognosis ($p < .0001$) [33]. In 41 patients with IPF who completed the Beck Depression Inventory (BDI), the mean score was 11.9 ± 6.0 and 8 (24%) subjects scored ≥ 15 [14]. Scores on this instrument range from 0-63; a score ≤ 9 is normal and the range of 10-18 suggests mild to moderate depression [23].

Although the stress of the diagnosis of a chronic medical illness typically induces anxiety and depression, these symptoms may diminish if there is hope for a cure. When the prognosis is unknown or the condition likely to be terminal, anxiety and depression can become more severe and prolonged. [34] Patients with IPF face challenges to their functional independence, self-esteem, and quality of life as a result of a progressive decrease in exercise capacity. Activities of daily living require extra planning and time. "Cough becomes a dreaded ordeal, and onset of exacerbations or new symptoms reawakens the fear of respiratory crisis or death" .[35] Faced with these stressors, patients with IPF are at high risk for developing anxiety and depression. These feelings may not be discussed during routine office visits. Because care providers typically focus on treatment options, prognosis, and relieving physical discomfort, they may not actively solicit concerns arising from anxiety and depressive symptoms from the patient or care providers. [34]

Studies have shown anxiety and panic are common in patients other types of lung disease, such as COPD and asthma, but little research has been done in patients with IPF. [35] The risk is that untreated anxiety or depression can further impact patients with severe, chronic medical illness and lead to risk of suicide, worsening of symptoms, delayed evaluation, and poor compliance [35] .While anxiety and depression are inevitable in patients with advanced lung diseases, both are highly responsive to treatment with medications and cognitive behavioral therapy [35].

2.3 COGNITIVE BEHAVIORAL THERAPY

One of the leading theories regarding the nature, cause, and treatment of depression is the cognitive behavioral therapy (CBT) developed primarily by Aaron T. Beck in the 1950's [36] . Beck views depression as "a multifaceted, biopsychosocial disorder characterized by negatively distorted cognitions and cognitive processes" [37]. This theory subscribes that depression is not mediated by stimuli within the environment, but rather the person's attendance to selected stimuli within the environment, [38], [39], [40] or their reaction based on previous experiences. This situation causes individuals to be predisposed to making arbitrary inferences based on insufficient evidence, taking information out of context, and placing blame on themselves, such as: "Bad things always happen to me" or "I brought this on myself".

According to Beck, in simplified terms, the cognitive model "states that dysfunctional disorders, psychiatric disorders, and psychological or behavioral disorders are characterized by dysfunctional thinking, and that the dysfunctional thinking accounts for the affective and behavioral symptoms" [39]. The major concepts that constitute this theory include negative automatic thoughts, dysfunctional attitudes, depressive schemata, and distorted thought processes [37].

The following schema presents an overview of Beck's theory.

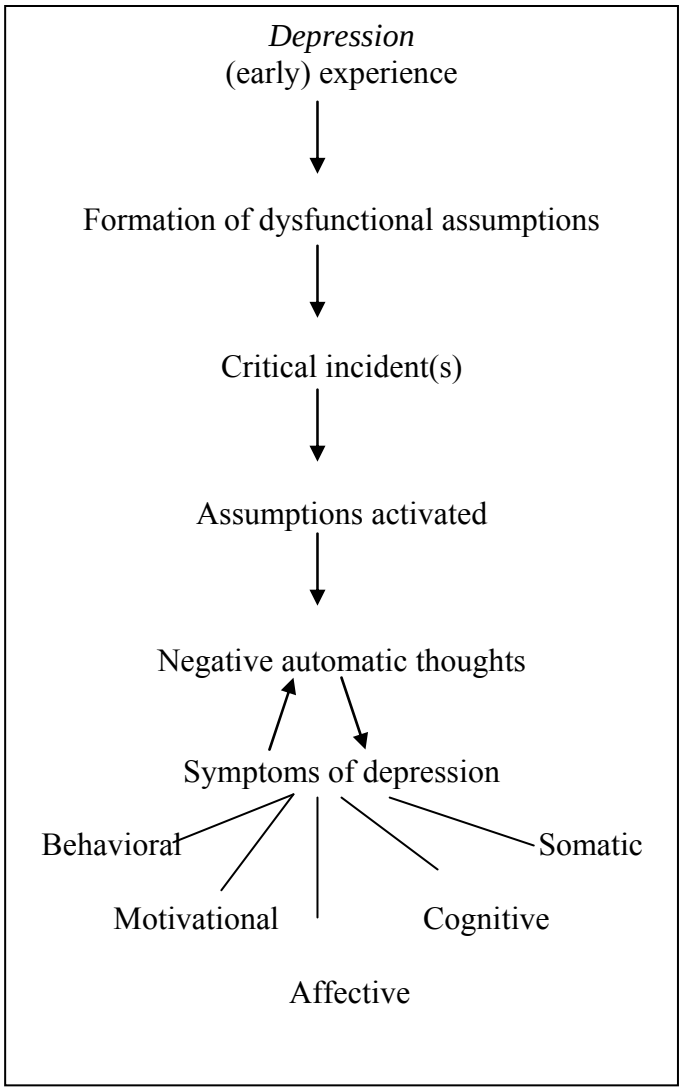

Figure 1. A cognitive model of depression.
(Adapted from: Houghton K, Salkovskis PM, Kirk J, Clark DM. Cognitive behavior therapy for psychiatric problems: a practical guide. Oxford: Oxford University Press, 1989:171). [39]

In his early work, Beck related that cognitive therapy "consists of all the approaches that alleviate psychological distress through the medium of correcting faulty conceptions and self-signals" [36] . Cognitive theory predicts that by modifying the patient's cognitive processes or how they think about a specific thing will help to reduce their depressive symptoms [41]. Beck also reported that cognitive therapy might be used during depressions to help the patient gain

objectivity toward his automatic reactions and counteract them. He added that during non-depressed periods, the therapy is designed to modify the idiosyncratic cognitive patterns to reduce the patient's vulnerability to future depression [42].

The benefits of cognitive theory are well supported in the literature [37], [41], [43], [44] and have gained widespread acceptance in the clinical community [45]. Scott contends that "there is considerable empirical support for the use of cognitive therapy in the treatment of mild to moderately severe acute major depression" [44]. His paper evaluated the role of cognitive therapy in day-to-day clinical practice, and concluded that cognitive therapy is the most widely researched brief psychological therapy for depression with over 80 randomized controlled trials to support its utility [45]. Whisman, in his review of cognitive therapy, adds that numerous studies have found cognitive therapy to be highly effective in the treatment for nonpsychotic, nonbipolar depression [46].

Cognitive behavioral interventions can benefit those who suffer from major depression, as well as those who experience depressive symptoms and anxiety [47], [48]. Patients with recurrent depression who were maintained at the same dose of their antidepressant medication, but who received six cognitive therapy sessions, had a lower relapse rate than those who only had their antidepressant medication dose increased and did not receive cognitive therapy [48]. The Program to Encourage Active, Rewarding Lives of Seniors (PEARLS) study, which tested a cognitive behavioral intervention in patients with minor depression, found that patients who enrolled in the program had a 50% reduction in depressive symptoms, 43% had complete remission from depression, and 36% had improved HRQoL (16). No studies were identified that studied the impact of cognitive behavioral therapy in patients with IPF.

2.4 HEALTH RELATED QUALITY OF LIFE (HRQOL).

If depression and anxiety are present, they are likely to impact HRQoL. Ratings of HRQoL provide a means of quantifying, in a standardized manner, how chronic illness impacts daily life, health and well-being [49], [50]. A limited literature has evaluated HRQoL in patients with IPF [14], [51], [52], [53],[54], [55], [56], [57].

In 1999, Chang, et al. at the University of Washington in Seattle, reported findings from a study they did to assess HRQoL in 50 patients with ILD from an outpatient pulmonary clinic in a university tertiary setting. The types of ILD included: IPF (n=33), sarcoidosis (n=10), and asbestosis (n=7). Four different quality-of-life instruments were administered to the patients: the Medical Outcomes Study Short Form 36 (SF-36), the Quality of Well-being scale (QWB), the Chronic Respiratory Questionnaire (CRQ), and the St. George's Respiratory Questionnaire (SGRQ). The patients also performed pulmonary function studies and a 6 minute walk [51]. Findings of this study showed that all four instruments were sensitive with pulmonary function study results, 6 minute walk, and dyspnea. The SF 36 and the SGRQ were the most sensitive and showed significance in relationship to the breathing results, 6 minute walk, and dyspnea.

Martinez, et al. evaluated HRQoL using the SF-36 questionnaire in 34 patients with IPF in a specialized outpatient respiratory clinic in Brazil compared with a group of 34 normal subjects. The SF-36 contains 8 components, including physical functioning, physical role, general health perceptions, pain, vitality, social functioning, emotional role, and mental health index. The patients with IPF scored significantly worse than the control subjects in seven of the eight components. The only component that did not show significance was the pain index. Martinez compared the SF-36 to the variables of the BDI and pulmonary function studies,

including arterial blood gases. The BDI score was significantly correlated with five of the eight components: physical functioning, general health perceptions, vitality, social functioning, and mental health index. Martinez demonstrated that the SF-36 was a valid instrument to measure quality of life in IPF patients [52].

DeVries, et al. studied HRQoL in patients with IPF. The aims of his study included assessing HRQoL and depressive symptoms, identifying factors related to HRQoL, and to identify impaired aspects of HRQoL to guide future rehabilitation programs. The patients completed the WHOQOL-100 (Dutch version) which is described as a cross-culturally developed, generic, multidimensional QoL measure, the BDI, and the Bath Breathlessness Scale. DeVries's findings suggested that HRQoL was mainly impair ed in the domains of physical health and level of independence when matched to healthy controls [14].

Swigris and colleagues interviewed 20 IPF pts to collect data on how IPF affects their lives, and identified 12 primary domains: symptoms, IPF therapy, sleep, exhaustion, forethought, employment and finances, dependence, family, sexual relations, social participation, mental and spiritual well-being, and mortality in an attempt to develop a disease specific instrument to measure HRQol in patients with IPF. Patients reported that IPF had a significant effect on their quality of life, especially with symptoms of breathlessness and cough being very bothersome, and that they had to rearrange their lives because of the disease. In addition, living with IPF made the patients reflect on their lives and their emotional aspects that affected them [54].

Jastrzebski, et al. evaluated HRQoL in 16 patients with IPF awaiting lung transplantation in Poland. He reported that scores on the SF-36 were significantly worse for physical functioning, role limitations due to physical problems and emotional limitations in everyday life.

One year later, he reevaluated the HRQoL of these patients and the role limitations due to physical problems were significantly worse [55].

Swigris and colleagues reviewed the 7 studies that evaluated HRQoL in IPF patients, and reported that patients with IPF were found to have impaired HRQL in nearly every life domain, but domains related to physical functioning, symptoms, and level of independence were affected most, with aspects of physical health being most negatively impacted [56].

Nishiyama and colleagues applied the Japanese version of the St. George Respiratory Questionnaire (SGRQ) to 41 patients with IPF to identify factors that significantly contribute to their HRQoL, and found that their dyspnea rating predicted their HRQoL more accurately than physiological variables, such as pulmonary function studies [57].

2.5 PERCEIVED STRESS.

Serious illness imposes substantial stress and burden on the patient and care partner. Prior studies indicate that caregivers of patients with disabling illness, such as cancer and dementia, have increased health problems and psychosocial stress [58], [59]. Findings from a study of nearly 1000 terminally ill patients and their caregivers [60] indicated that patients who have substantial care needs report that they have a subjective sense of economic burden. In addition, caregivers of these patients are more likely to have a high level of stress and depressive symptoms [61]. Several studies have demonstrated that a psycho-educational intervention can benefit those who have serious illness accompanied by stress and depression and can improve their psychological well-being [62], [63], [64], [65]. While much of the literature focuses on those who experience a long-term illness trajectory and their caregivers, the psychosocial impact of a psycho-educational

intervention targeted early in the onset of a potentially life ending illness, such as IPF, has not been examined.

2.6 DISEASE MANAGEMENT INTERVENTION.

Important aspects of well-being may be adversely affected by the diagnosis of a progressive, ultimately fatal illness [33]. While progression may be rapid, the more common scenario is a gradual decline in function, accompanied by increasing disability. During this interval, a primary goal is to assist patients to live life as fully as possible by reducing symptom burden, maintaining functional ability, and thereby achieving more optimal HRQoL [66].

Several approaches may be used to achieve this goal. *Palliative care*, a relatively new discipline in healthcare, is the active care of patients whose disease is not responsive to curative treatment [67], [68]. The goal is to provide care that enhances patient comfort and function from diagnosis, through treatment, and the terminal phase and to also support the patient's care partner [67]. Initially used primarily for patients with cancer, palliative care is increasingly recognized as relevant to the care of advanced disease in a variety of settings [67]. Whenever possible, the transition to palliative care should be planned in advance for patients with progressive, irreversible lung disease, such as IPF. For more than 20 years, the advanced directive has served as the primary tool in planning care at the end-of-life[17].

Advanced care planning is the process of reflection, discussion, and communication of treatment preferences for end-of-life care. The process of advanced care planning begins with the patient's understanding of the disease process and talking about the possible scenarios that may lead to death in hope that the patient will be better prepared for what to expect as the disease

progresses and what options may be available [16]. Advanced care planning promotes better end-of-life care as patients participate in decisions that affect his/her own future. Advanced planning helps the patient become more autonomous and promotes self-determinism [16].

Cognitive behavioral therapy has been shown to benefit those who suffer from major depression, as well as those who experience depressive symptoms and anxiety [47], [48]. The benefits of cognitive theory are well supported in the literature [37], [45], and have gained widespread acceptance in the clinical community [45]. Scott contends that "there is considerable empirical support for the use of cognitive therapy in the treatment of mild to moderately severe acute major depression" [44].

Pulmonary rehabilitation programs, commonly used to manage symptoms of patients with chronic obstructive lung disease (COPD), seek to reduce symptoms and improve functional ability in patients with disabling dyspnea. Studies of the efficacy of pulmonary rehabilitation in IPF have not been done.

While each of these approaches has been shown to be beneficial, the concerns of patients with a progressive, ultimately fatal condition are likely to be multifocal. Accordingly, they may be more optimally managed by a *disease management intervention* that addresses the multiple problems that such individuals may be encountering. The proposed intervention is designed to be brief, a necessary requirement to be feasible in this clinical setting, but comprehensively address primary patient concerns. The 6 sessions will address: 1) diagnosis and treatment of IPF; 2) strategies to manage moods and feelings; 3) strategies to manage anxiety and depressive symptoms; 4) advanced care planning; 5) strategies to manage symptoms and promote exercise; and 6) a wrap-up and synthesis session. Each will include an educational session, discussion, and homework related to the topic.

The purpose of this study will be to determine if patients with IPF and their care partners could be more optimally managed by a *disease management intervention* that nurses can deliver using the format of a support group which addresses the multiple problems that such individuals may be encountering. A schema of the conceptual model of this study (Figure 2) follows:

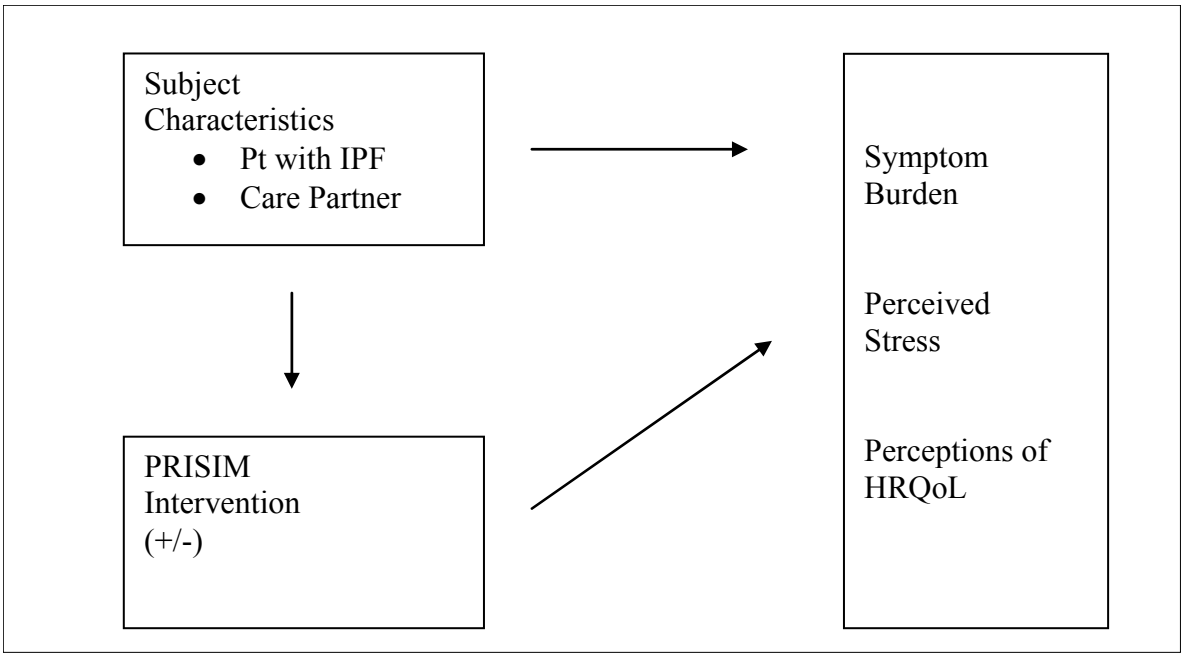

Figure 2. Conceptual Model.

2.7 SUMMARY

The prognosis for IPF is poor [5] , [8], [9]. In most patients, symptoms have been present for more than 6 months before they present to their doctor. The clinical course is one of gradual deterioration, but occasionally there are periods of rapid decline with this disease, which often represent accelerated disease. The median survival for these patients is 3-5 years with the

majority of deaths occurring from progressive respiratory failure [6]. As the disease progresses, the patients become more symptomatic with shortness of breath and cough, more limited in their functional status, and dependent upon supplemental oxygen [10]. Considering this poor prognosis, it is important to evaluate the symptom burden, including anxiety and depression, perceived stress, and quality of life of the patient with IPF and their care partner. The goals of treatment of IPF include managing symptoms and relieving discomfort. Palliative care should be offered from the time of diagnosis [7].

The use of an intervention, such as PRISIM, that includes information on various aspects of palliative care, including scenarios dealing with advanced planning, exercises incorporating cognitive behavior therapy techniques, and how to live with the disease despite the uncertainty of the disease course via different pulmonary rehabilitation aspects of symptom management and exercise, may affect the impact the disease has on the patient and care partner. A mixed methods approach using both the quantitative and qualitative measures should provide a comprehensive description of the ability of a *disease management intervention* (PRISIM: Program to Reduce Symptoms and Improve Lifestyle Management) to decrease symptom burden, decrease perceived stress, and improve perceptions of health-related (HRQoL) for patients with IPF and their care partners.

3.0 PRELIMINARY WORK

This chapter reviews the preliminary work that was conducted to assess the disease impact of IPF and provide pilot work for the dissertation study.

3.1 PURPOSE

The purpose of this pilot study was to determine the self-reported prevalence of anxiety and depressive symptoms in patients with IPF and examine relationships between these psychological variables and HRQoL, sleep disturbances and social support.

3.2 METHODS

3.2.1 Site & Sample.

The sample consisted of 28 patients with IPF who were recruited from a specialized university-based adult center for the care of patients with interstitial lung disease. To enter the study, subjects had to be: 1) diagnosed with IPF on the basis of biopsy or clinical (high resolution computed tomography [HRCT]) findings; 2) forced vital capacity (FVC) reflecting moderate

(FRC ≥ 55- 70 % pred) or severe (FVC < 55 % pred) disease; 3) ≥ 18 years of age; and 4) able to speak and write English. To insure a broad range of disease, recruitment was targeted to achieve a sample that was stratified to include equal numbers (50%)of patients with moderate and severe disease based on the most recent FVC recorded in the medical record. The study was approved by the University of Pittsburgh Institutional Review Board and all subjects provided informed consent.

3.2.2 Measures.

Anxiety was measured using the Spielberger State-Trait Anxiety Inventory (STAI) which consists of two 20-item forms that measure general (trait) and current (state) anxiety. The two forms are scored by adding weighted scores for each item. Scores range from 20 – 80 with higher scores indicating greater anxiety [69]. *Depression* was measured using the *Beck Depression Inventory-II (BDI),* a 21-item self-report instrument designed to measure the severity of depressive symptoms in adults and adolescents 13 years or older. Scores range from 0 to 63. Scores of 5-9 indicate no or minimal depression, 10 – 18 indicate mild to moderate depression, 19 – 29 indicate moderate to severe depression, and 30 – 63 indicates severe depression [70]. *HRQoL* was measured using the Medical Outcomes Study Short Form 36 version 1 (SF-36), a 36-item widely used generic scale used to assess HRQoL in a variety of chronic medical conditions [25] and validated in patients with IPF [52]. Scores on 8 domains can be grouped into a physical component score that includes scores for physical functioning, role physical, bodily pain, and general health and a mental component score that includes scores for vitality, social functioning, role emotional, and mental health. Lower scores indicate less positive perceptions of HRQoL. *Sleep disturbances* were measured using the Pittsburgh Sleep Quality Index (PSQI), a

self-rated questionnaire that assesses sleep quality and disturbances during the previous month [71]. The PSQI contains 19 items which generate 7 component scores: subjective sleep quality, sleep latency, sleep duration, habitual sleep efficiency, sleep disturbances, use of sleeping medication and daytime dysfunction. Scores are summed for the 7 components to yield a global score (range 0-21). Higher scores indicate more impaired sleep quality. *Social Support* was measured using the Interpersonal Support Evaluation List (ISEL) [72]. The ISEL was designed to assess the perceived availability of social support and provide an overall measure of support. The ISEL consists of 40 items that fall into four 10-item subscales designed to measure perceived availability of: 1) material aid (tangible); 2) someone to talk to (appraisal); 3) positive comparison to self (self-esteem); and 4) people to do things with (belonging). The total score ranges from 0–160. Higher scores indicate greater support. Demographic data were obtained from the subject's medical record.

3.2.3 Procedures.

Patients were contacted by in person or via telephone by clinic personnel and, if willing to participate, given or mailed a copy of the informed consent, questionnaires, and a self-addressed return envelope.

3.2.4 Statistical Analysis.

Data were analyzed using descriptive statistics, Chi-square, t-tests, ANOVA, and Pearson's correlation coefficients, as appropriate. All statistical analyses were performed using SPSS software.

3.3 RESULTS

3.3.1 Demographics.

The sample consisted of 28 patients (53.6% male) who were 67.8 ± 8.4 years of age (Table 1).

Table 1. Demographic and Clinical Characteristics (N = 28)

Variable*	Total Group	Moderate (n=14) (FVC ≥ 55% - 70%)	Severe (n=14) (FVC < 55%)	p
Age, yrs	67.8 ± 8.4	68.9 ± 9.4	66.7 ± 7.5	.51
Gender, % male	53.6%	57.1%	50.0 %	.70
Education, yrs	14.5 ± 3.2	15.3 ± 3.6	13.6 ± 2.8	.19
Smoking History, % never	32.1	21.4%	42.9%	.41
Diagnosis				
Biopsy, %	57.1%	50%	64.3%	-
Clinical (HRCT)	42.9%	50%	35.7%	-
Lung Function				
FEV_1, % pred	61.8 ± 19.2	69.9 ± 22.7	53.7 ± 10.6	.03
FVC, % pred	53.5 ± 13.3	64.2 ± 8.0	42.8 ± 7.4	<.001
FEV_1FVC % pred	86.8 ± 8.5	84.6 ± 7.97	89.0 ± 8.8	.19
DLCO, % pred	39.1 ± 11.8 ^	42.6 ± 10.3	35.3 ± 12.5	.11
Oxygen use, % yes	21.4%	21.4%	21.4%	.64
Oral steroid use, % use	57.1%	28.6 %	71.4%	.007
Immunosuppressives				
Actimmune, %	39.3%	35.7%	50.0%	.44
Cytoxan, %	7.1%	0%	14.3%	-
Immuran, %	7.1%	0%	14..3%	-
Psychotropics, % yes	18.5%^	23.1%	14.3%	.92

* mean ± SD unless indicated; ^ n=27

Mean FVC was 53.5% ± 13.3%. Diagnosis of IPF was confirmed by biopsy in 57.1% and high resolution computed tomography (HRCT) in the remainder. While there is no known treatment for IPF, 39.3% of the overall sample reported taking interferon gamma injections,

known as Actimmune, which increased to 50% (n=7) in patients with severe disease A minority with severe disease reported taking azathioprine (Imuran) (14.3%) or cyclophosphamide (Cytoxan) (14.3%). More patients with severe disease reported use of oral steroid therapy (p=.007) compared to patients with moderate disease. There were no significant between group differences in regard to any other demographic variable or clinical characteristic with the exception of FEV and FVC% predicted, as anticipated given sample characteristics.

3.3.2 Disease Impact

Mean scores on the trait form of the STAI indicated more anxiety in the patients with severe disease, compared to those with moderate disease approaching statistical significance (p = 0.06) (Table 2).

Table 2. Anxiety, Depressive Symptoms & HRQoL (N = 28)

Variable	All Subjects		FVC%<55%		FVC%>55%		
	Mean	SD	Mean	SD	Mean	SD	**p**
STAI trait	54.00	5.46	52.1	4.9	55.9	5.5	**0.06**
BDI tot	14.10	10.60	14.0	11.6	14.2	9.9	0.96
SF-36 Tot	80.22	16.30	79.8	18.5	80.6	14.4	0.89
SF-36 P	28.68	10.96	29.1	11.3	28.3	11.0	0.86
SF-36 M	51.54	13.11	50.7	14.2	52.3	12.4	0.75
PSQI tot	7.89	3.93	6.8	2.7	9.0	4.7	0.14
ISEL Tot	89.43	22.91	93.0	22.4	85.9	23.7	0.42

*mean ± SD unless indicated

Scores on the BDI were equivalent for those with moderate and severe disease (P=.96) with a wide variation for the total group (range 2-48). Of the 28 subjects, more than 50% of the sample reported some degree of depressive symptoms, with 36% reporting mild - moderate depressive symptoms, 11% reporting moderate - severe depressive symptoms, and 11% reporting severe depressive symptoms (Figure 3).

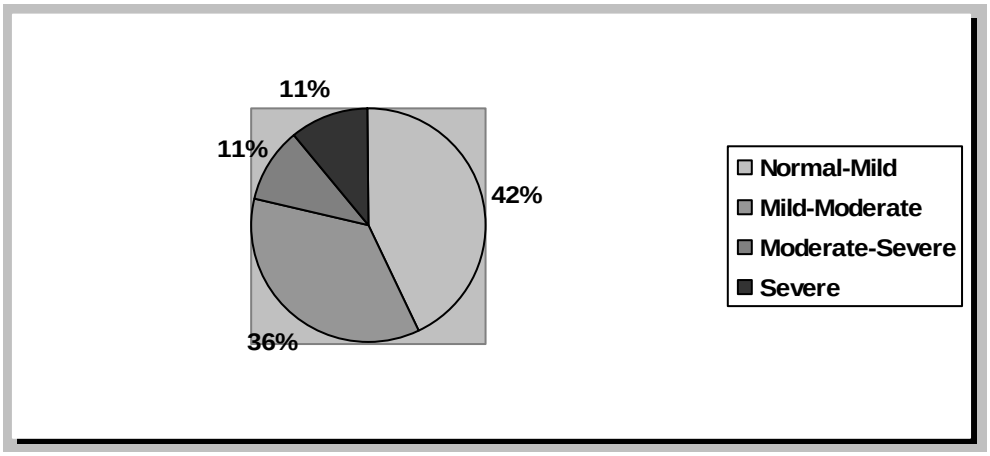

Figure 3. BDI Scores grouped by level of severity

Five (18.5%) subjects were prescribed psychotropic medications. Further review of those five subjects revealed higher scores on the BDI; two in the mild-moderate range, two in the moderate-severe range, and one in the severe range. As required by the IRB, subjects whose scores indicated severe depressive symptoms were contacted by the research team and their primary care provider was also notified. They were both informed of the findings, and further assessment was advised, as well as referrals to mental health resources.

Patients scored lower on the physical component of the HRQoL (28.68 ± 10.96) in comparison to the mental component (51.54 ± 13.11) indicating less satisfaction with their quality of life, especially in the areas of physical function, role physical and general health. The

physical component and mental component scores can be normalized to responses from the general population where the mean score is 50 (Figure 4). When viewed from this perspective, all scores were less than norms for the general population with the total mental score very close to the norm, while the total physical scores were much lower.

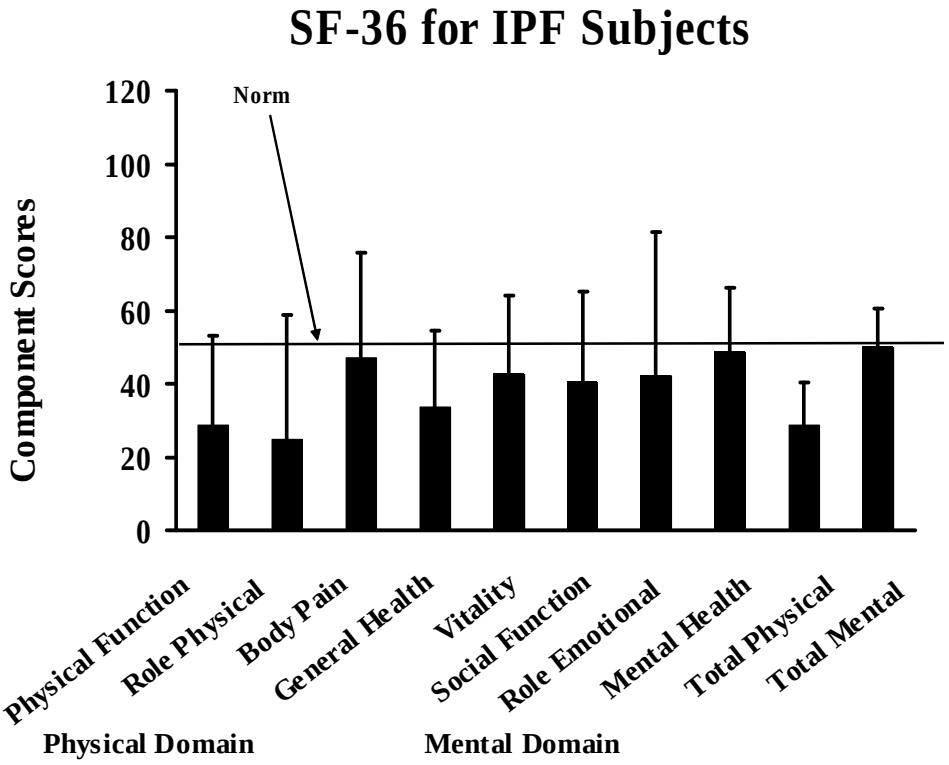

Figure 4. : HRQoL Scores Normalized to General Population Scores

The mean PSQI global sleep quality score was 7.89 ± 3.93 for the total sample, which is approximately midway between scores for a normative sample of healthy "good" sleepers and for "poor" sleepers with a major depressive disorder, respectively [71]. The mean score for the ISEL was 89.43 ± 29.7. Higher scores on the ISEL indicate higher support [72].

3.3.3 Relationships between Measures.

Lower scores on the SF-36 (less positive ratings of HRQoL) were associated with perceptions of less social support, and more depressive symptoms ($p < .01$) (Table 3).

Table 3. Pearson Correlation Coefficients between measures of disease severity, anxiety, depression, HRQoL, social support, sleep, dyspnea, gender and age. (N = 28)

	Gender	Age	Time p Dx	FVC %	SF36 Tot	SF36 P	SF36 M	ISEL Total	BDI Tot	STAI Trait	PSQI Tot	SOBQ
Gender	1											
Age	.283	1										
Time p Dx	.244	.147	1									
FVC%	.118	.051	-.160	1								
SF-36 Tot	**-.427(*)**	-.030	-.114	.080	1							
SF-36 P	-.020	.014	-.099	.055	.599(**)	1						
SF-36 M	**-.513(**)**	-.049	-.058	.054	.743(**)	-.091	1					
ISEL Tot	-.368	-.014	-.138	-.046	**.656(**)**	.103	**.729(**)**	1				
BDI Tot	**.515(**)**	-.019	-.072	-.162	**-.715(**)**	-.196	**-.726(**)**	**-.695(**)**	1			
STAI Trait	**-.374(*)**	-.264	-.330	.368	.003	-.107	.093	.007	-.195	1		
PSQI Tot	.248	-.051	.060	.299	-.183	.099	-.310	-.310	.195	-.215	1	
SOBQ	-.083	-.054	-.124	.053	.071	-.373	**.400(*)**	-.051	-.036	.167	-.075	1

** Correlation is significant at the 0.01 level (2-tailed).
* Correlation is significant at the 0.05 level (2-tailed).

Lower scores on the SF-36 mental component scores (less positive perception of HRQoL) were associated with more depressive symptoms ($p <.01$) and dyspnea ($p < .05$). Perceptions of less social support were associated with more depressive symptoms, and a less

positive perception of HRQoL as determined by the total SF-36 score and the SF-36 mental component score ($p < .01$). No significant correlations were seen for sleep, age, time after diagnosis, or disease severity.

Although not a focus of this study, the data also revealed significant gender-based differences (Tables 3 and 4). Men rated their HRQoL (SF-36 mental component score) more positively than women ($p = .005$). Men rated their anxiety higher ($p = .05$) than women but reported fewer depressive symptoms ($P = .005$) than women.

Table 4. Gender Ratings of Depressive Symptoms and Anxiety (N=28)

	Gender	N	Mean	Std. Deviation	p
BDI	Men	15	9.1	5.7	**.005****
	Women	13	19.8	12.1	
STAI	Men	15	55.9	5.0	**.05***
	Women	13	51.8	5.4	
SF36 Total	Men	15	86.57	16.14	**.024***
	Women	13	72.88	13.60	
SF36 P	Men	15	28.88	14.38	.921
	Women	13	28.45	5.36	
SF36 M	Men	15	57.7	7.42	**.005****
	Women	13	44.42	14.86	

*p < .05. **p < .01.

3.4 DISCUSSION

The purpose of this pilot study was to determine the self-reported prevalence of anxiety and depressive symptoms in patients with IPF and examine relationships between these psychological variables and HRQoL, sleep disturbances and social support. An important finding

was that greater than 50% reported anxiety and depressive symptoms, decreased HRQoL, and the lack of social support.

As might be expected, individuals who perceived their HRQoL as less positive also perceived themselves as having less social support and reported more depressive symptoms. In addition, these individuals reported more dyspnea, which may have influenced ratings of HRQoL. The perception of more social support seemed to have beneficial effects, as those patients reporting less social support also reported more depressive symptoms and a less positive perception of their HRQoL. There was a positive association between the dyspnea scores on the SOBQ score and the mental component of the SF-36 indicating that patients with increased dyspnea rated their HRQoL lower in areas of vitality, social function, and role emotional areas of the mental component score of the SF-36.

Sleep, age, time after diagnosis, and disease severity did not have any significant impact on the psychological variables. This was surprising as depressive symptoms can influence sleep quality.

While this was not an aim of this study, findings indicated significant gender differences between women and men in self-reported anxiety, depression, and perceptions of HRQoL. Men reported fewer depressive symptoms but more anxiety.

Findings from this study are similar to those of Kunik and colleagues in a study of 1,334 patients with chronic breathing disorders [73]. When screened for anxiety and depression, 34 – 72% of patients were found to have anxiety and/or depression [73]. An interesting additional finding of their study was that <40% of patients with COPD were being screened for anxiety/depression. In studies of patients with other respiratory disorders, depression has also been reported to be common and persistent, e.g., patients with lung cancer [33]. In the sample of

patients awaiting lung transplantation, 25% had a diagnosis of at least one mood or anxiety disorder, including depressive symptoms and panic or anxiety disorder [74].

Findings of this study regarding the relationship between depressive symptoms and a lower ranking of HRQoL are similar to those of De Vries and colleagues who reported that 24% of patients with IPF scored in a range suggesting mild to moderate depression and also reported a more negative perception of their HRQoL [14].

Given the large proportion of patients who report depressive symptoms, findings suggest the need to query patients regarding their psychological response to their illness and attempt to identify individuals for treatment to improve management of depression. Others have advocated a more active search as a means to better manage depression in primary care [75]. Although no cure exists for IPF, prompt detection of anxiety and depression and appropriate treatment may assist patients to more optimally cope with the consequence of this illness and its impact on their HRQoL

3.5 CONCLUSION

Depression, anxiety, and lower reported HRQoL were found to be prevalent in patients with IPF. Social support was found to potentially mediate the impact of IPF on these symptoms. While the sample characteristics were reflective of the served population, it was small in size. The standard of care for patients with IPF should include assessment of psychosocial factors that may impact on the patient's overall quality of life. Referral for appropriate management can then be implemented, based on individual responses.

4.0 METHODOLOGY

This study will use two modes of inquiry to test the ability of a *symptom management intervention* (PRISIM: Program to Reduce Symptoms and Improve Lifestyle Management) to decrease symptom burden, decrease perceived stress, and improve perceptions of HRQoL for patients with IPF and their care partners. The quantitative and qualitative inquiries will be conducted in order.

4.1 QUANTITATIVE INQUIRY

The purpose of the quantitative portion of the study will be test outcomes of providing a structured 6-session educational program to patients and care partners of patients with IPF. The sessions will be structured to include disease-specific information as well as information potentially helpful in coping with the illness.

4.1.1 Research Design.

This goal of this study is to enroll 24 patients and 24 caregivers. Twelve patients and 12 caregivers will be randomized to an intervention group and 12 patients and 12 caregivers to a usual care group. Patients in the intervention and usual care group will complete questionnaires

designed to assess shortness of breath, anxiety, depression, perceived stress, and HRQoL the week before study entry (baseline) and one week after completion of a 6 week intervention program (week 7). Caregivers will complete questionnaires designed to assess anxiety, depression, HRQoL, and perceived stress at the same intervals (Table 5). In prior work, the total time required to complete the questionnaire battery was approximately one hour.

Table 5. Study Design

Time	Study Entry	Weeks 1-6	Week 7	Week 26
Group I	Questionnaire Battery	Intervention	Questionnaire Battery	Questionnaire Battery
Group II	Questionnaire Battery	Usual Care	Questionnaire Battery	Questionnaire Battery

4.1.2 Setting and Sample.

Patients will be recruited from the Simmons Center for Interstitial Lung Disease located in the Comprehensive Lung Center, UPMC-MUH. To be eligible, patients have to be: 1) 21 years or older; 2) able to read and understand English; 3) diagnosed with IPF; and 4) have a FVC that reflects moderate or severe disease. Care partners, e.g. an individual who lives with and/or care for the patient will have to be: 1) 21 years or older; 2) able to read and understand English; and 3) live with and/or care for the patient with IPF who is eligible for inclusion. To enroll, both the patient and the care partner will have to consent to study participation. Children will not be included because the disease condition being examined typically does not occur until 40 years of

age. The racial, gender and ethnic characteristics of the proposed subject population is anticipated to reflect the demographics of Pittsburgh and the surrounding area and/or the patient population currently seen in the Simmons Center for Interstitial Lung Disease. We shall attempt to recruit subjects in respective proportion to these demographics. No exclusion criteria shall be based on race, ethnicity, gender or HIV status.

4.1.3 Rationale for Sample Size.

NCSS/PASS 2004 software was used to determine the estimated sample size: Sample size was determined by power analysis for the PRISIM study. A total sample size of 18 patients plus 18 care partners in each group (experimental and control) to equal a total of 36 dyads is recommended to attain a power of >0.80 with an alpha level of 0.05. Given the availability of patients in the Simmons Center, the sample will be a convenience sample defined as the first 24 subjects who meet the inclusion criteria, give consent to participate, and participate in the study by answering all the questions.

4.1.4 Measurement of Variables.

Five dependent variables will be measured and analyzed. They are anxiety, depressive symptoms, stress, and HRQoL for both the patient and caregiver. Shortness of breath will also be measured for the patient with IPF. The level of measurement for each of these five dependent variables represents interval data. The independent variable is the PRISIM intervention and this represents nominal data. The stratifying factor will be the type of member; the patient with IPF or their care partner (nominal data). In addition, other stratifying factors include the severity of

disease measured via FVC (interval data) for the patient, and whether the patient and their care partner previously participated in the IPF support group (nominal data).

4.1.5 Measures.

Demographic & medical profile data will be obtained from the patient and from the care partner (Appendix A). The demographic information, to be used descriptively, includes: age, gender, race, education (in years), diagnostic method (open lung biopsy or high resolution computerized tomography (HRCT) scan, employment status, annual income, length of time since diagnosis (in months), concurrent medical problems, smoking status, medications for IPF, and prior or current therapy for depression. The following objective measures will be obtained from their most recent clinic visit: spirometry (FVC, forced expiratory volume in one second (FEV_1), diffusion capacity (DLCO), and exercise desaturation test results.

Dyspnea will be measured using the *University of California at San Diego Shortness of Breath Questionnaire (SOBQ)* (Appendix B). The SOBQ is completed by asking subjects to rate the severity of shortness of breath on a 6-point scale (*0=not at all to 5=maximal or unable to do because of breathlessness*) during 21 activities of daily living (ADL's) associated with varying levels of exertion. There are 3 additional questions that ask about daily life limitations due to shortness of breath, fear of over exertion and fear of shortness of breath. If subjects do not routinely perform the activity, they will be asked to estimate the shortness of breath anticipated. The score is obtained by summing responses on the 24 items to form a total score (range 0-120). In psychometric testing, internal consistency was $\alpha = 0.96$ [21]. Item-total corrections ranged from 0.49 to 0.87 (28). SOBQ scores were negatively correlated with physiologic measures of disease severity (FEV_1, DCO), HRQoL (Quality of Well-Being) and exercise tolerance (6-

Minute Walk) (r= –0.41 to -0.68) and positively correlated with ratings of perceived breathlessness (Borg Scale) and depression (Center for Epidemiological Studies-Depression Questionnaire) (r= 0.37 to 0.45) [21].

Anxiety will be measured using *The Beck Anxiety Inventory (BAI)* (Appendix C), a standardized, 21-item tool designed to assess the presence and severity of common symptoms of anxiety, both psychological (e.g., feeling nervous, scared) and physical (e.g., heart pounding, hands trembling). The tool uses a 4-point scale (0 = absent/not at all disturbing to 3 = I could barely stand it). Items are summed and total scores range from 0 to 63. A scores of 0 to 7 indicates no anxiety, 8 to 15 mild anxiety, 16 to 25 moderate anxiety, and scores 26 or higher, severe anxiety [76]. Reported mean Beck Anxiety Inventory scores for psychiatric outpatients with Diagnostic and Statistical Manual of Mental Disorders, Revised Third Edition diagnoses of anxiety disorders range from 17 to 29 [77]. Means for community samples have ranged from 7 to 11 [78], [79].

Depressive symptoms will be measured using the *Beck Depression Inventory-II (BDI)* (Appendix D). The BDI-II, a 21-item self-report instrument, is designed to measure the severity of depression in adults and adolescents 13 years or older [80]. The BDI-II, to be used in this study, is a revised version of the original instrument. A meta-analysis of studies reporting psychometric properties of the BDI-II cites advantages in terms of content validity and ability to discriminate between depressed and non-depressed people [81]. Internal consistency ranges from 0.73 to 0.92 with a mean of 0.86 [23]. Correlations with clinician ratings of depression using the BDI-II ranged from 0.62 to 0.66 [82]. Scores range from 0 to 63. A score of 0-13 suggests minimal depression, 14-19 mild depression, 20-28 moderate depression and 29-63 severe depression [81].

Perceived Stress will be measured by the Perceived Stress Scale (PSS) [72]. The PSS (Appendix E) is designed to measure the degree of perceived stress of a situation, and the degree to which subjects find their lives unpredictable, uncontrollable, and overloading. Individuals are asked to indicate how they feel or thought in a certain way in the last month using the 5 options, 0 (never) to 4 (very often). The range of scores is from 0 to 40, with higher scores indicating more appraised stress. There are no cut-off points on the PSS. The internal consistency and validity have been well established with a diverse population [83], [84], [85].

HRQoL will be measured using the *SF-36* (Short Form Medical Outcomes Study Form) Version 2 (Appendix F), a widely used generic scale, that has been used to assess HRQoL in a variety of chronic medical conditions and validated in patients with IPF [47, 52]. The SF-36 assesses 8 domains of HRQoL. Scores range from 0 = maximum impairment to 100 = no impairment. Scores on the 8 domains can be grouped into a physical component score (PCS) that includes scores for physical functioning, role physical, bodily pain, and general health and a mental component score (MCS) that includes scores for vitality, social functioning, role emotional, and mental health [86]. The PCS and MCS can be normalized to responses from the general population (mean score is 50). The validity, reproducibility, and responsiveness of the SF-36 have been well-demonstrated [87].

4.1.6 Procedures

The study will be conducted at the Comprehensive Lung Center where the research staff is part of the treatment team. Potential patients and their care partners will be first identified by their clinical care team who are also members of the research team. Research personnel responsible for administering the questionnaires will be blinded to group. The clinician will discuss the

research project with the patient and care partner. If they express interest in study participation, the clinician will explain the study in further detail and obtain informed consent. After consent is obtained, a medical record review of the patient's chart for inclusion/exclusion criteria will be conducted. A notation will be made in the progress notes of the subject's chart disclosing this review. If the patient does not meet entry criteria, he/she and the care partner will be removed from the study

Patients/care partners who provide informed consent will be assigned to group using cards prepared by an individual not associated with the study and placed in numbered opaque envelopes. The envelope will be opened and the patient informed of group designation after informed consent is obtained.

The intervention will consist of a 6 session symptom management intervention. Each session will be 2 hours in length and take place at UPMC Presbyterian in the Scaife Conference Center. The sessions included:

Session 1: *What is IPF and How to Live with it?* will review the causes, pathophysiology, and treatment of IPF. Participants will also receive a copy of the book, "Feeling Good" by Dr. David D. Burns that will be used for assignments at Session 2 and 3, and read by the patient or care partner at their leisure [88].

Session 2: *Gaining Control of Your Moods and Feelings: You Feel the Way You Think* will begin with a discussion of the basic principles of cognitive behavior techniques including the negative triad and the role of automatic thoughts. Ten cognitive distortions will be discussed and group members will be prompted to share examples of how they use distortions in their own lives. Group members will be encouraged to discuss these distortions with other group members

by giving concrete examples related to their lung disease. Short homework activities will be assigned.

Session 3: *Gaining Control of Your Moods and Feelings: What Can You Do About Depression* will begin with the group asking members to share examples from their homework. Members will be encouraged to provide feedback to the other group members. The group leader will discuss the concepts of stress and depression, and how they may relate to the group members and their care partners. Treatment modalities for depression and benefits gained from effectively dealing with depression will be discussed.

Session 4: *Putting Your Life in Order: What Do I Do Now?* will begin with group members being asked to share their observations about life and planning for uncertainty while trying to maintain control. The group leader will address important end-of-life issues including struggling with the reality of terminal illness, working with the medical team, helping family and loved ones cope with the illness, and planning ahead to set their affairs in order.

Session 5: *Living with IPF* will focus on living with IPF. Using lecture and group discussion, symptom management, energy conservation, correct use of oxygen therapy, and the importance of exercise will be addressed.

Session 6: *Wrap-up and Review Group* will encourage group members to review what they have learned during the past sessions and how they could apply this to their lives.

Group sessions will be led by nurses who are trained as a pulmonary clinical nurse specialist (Session 1, 5, & 6), advanced care planning instructor (Session 4), and a psychiatric clinical nurse specialist with additional training as a cognitive behavioral therapist (Session 2 & 3). Concurrent with this study, subjects will receive their usual medical care.

Control subjects will receive usual medical care during the study. If they request, the 6 week intervention will be presented after completion of the study..

4.1.7 Data Analysis

Demographic data will be collected on both the subjects and caregivers. Descriptive statistics will be computed by treatment group, type of member, and time. Study data will be analyzed to address the specific aims using descriptive statistics and MANOVA as appropriate. For all statistical tests, a p-value of less than 0.05 will be used to indicate statistical significance.

4.2 QUALITATIVE INQUIRY

The purpose of the qualitative portion of this mixed methods approach will be to generate an explanatory theory of the experiences of the patients and their caregivers in the PRISIM groups, using grounded theory methodology [89], [90]. Grounded theory is guided by symbolic interaction, a philosophical foundation described by Blumer that purports that people construct meaning for phenomena based on the interpretations of the interactions that they have with one another [91]. It is through social interactions that persons construct meaning. In the case of the patients and their care providers, this study seeks to understand the meaning they may construct for and the experiences they will have in the PRISIM groups.

4.2.1 Research Design

This study will enroll patients and caregivers from the experimental group. Each patient and care partner will be interviewed individually to elicit information on their experience after participating in PRISIM.

4.2.2 Setting and Sample

Each patient and care partner will be interviewed in the home setting approximately 3-9 months after participation in the intervention. By observing the patient and care partner in the home setting, it will give the researcher the ability to see the patient in the naturalistic setting and see their daily life experiences. Data collection will occur through open-ended questions (see Attachment 2). Responses to the questions will guide future questions and become more focused as the interviews progress.

4.2.3 Rationale for Sample Size

Sample size will be determined by the number of interviews necessary to achieve theoretical saturation, a situation in which no new information is elicited through data collection.

4.2.4 Procedure

Verbatim transcripts will be analyzed line by line initially and then more generally by comparing transcripts to transcripts. Weekly meetings with a faculty expert in qualitative research will be held to debrief about ongoing data analysis. Grounded theory will guide data analysis.

4.2.5 Data Analysis

Data analysis will consist of open coding, wherein codes or concepts will be developed that reflect the meaning in the transcripts. During open coding the researchers are very inductive and open to any and all codes that possibly reflect the meaning in the data. These codes are then analyzed through a process referred to as axial coding, in which the complexities of each code are explored. For example, the codes are analyzed according to such variables as under what conditions the codes occur, the consequences of these codes. Eventually, only certain codes are supported in the ongoing data analysis and those codes are then looked at in even more depth through process called selected coding. Those selective codes are then integrated into an explanatory framework to reflect the overall meaning in the data.

5.0 RESULTS

The results of the study were submitted in the form of a manuscript.

5.1 ABSTRACT

Palliative care is increasingly recognized as relevant to the care of advanced disease in a variety of settings. Idiopathic pulmonary fibrosis (IPF) results in scarring of the lung, respiratory failure and has a median survival of 3-5 years from time of diagnosis. The purpose of this study was to evaluate the impact of a 6-week program designed using palliative care concepts on symptom burden and health-related quality of life (HRQoL) in patients with IPF and their care partners. We hypothesized that participation would improve perceptions of HRQoL and decrease symptom burden.

Subjects were 42 participants randomized to an experimental (10 patient/care partner dyads) or control (11 patient/care partner dyads) group. Experimental group participants attended the 6-week program and controls received usual care. Prior to and after the program, all participants completed questionnaires designed to assess anxiety, depression, perceived stress, and HRQoL.

Following the intervention, experimental group patients rated their HRQoL less positively (p=.038) and tended to report more anxiety (p=.077) compared to controls. Care

partners rated their stress at a lower level (p=.018) compared to controls. Course evaluations were uniformly positive. Post study qualitative interviews with experimental group participants suggested benefits not exemplified by these scores. Patient participants felt less isolated, were able to put their disease into perspective, and valued participating in research and helping others.

Further exploration of the impact of palliative interventions in patients with advanced lung disease and their care partners is needed using both qualitative and quantitative methodology.

5.2 INTRODUCTION

Idiopathic Pulmonary Fibrosis (IPF), the clinical condition characterized by progressive dyspnea and chronic cough, restrictive lung disease, and the histopathologic pattern of usual interstitial pneumonia (UIP) [5, 92, 93], is one of the most common forms of interstitial lung disease [94] . IPF is often thought of as a disease with uniformly poor survival, but the clinical course varies widely [7, 95]. Some patients die within one year of diagnosis, whereas others live longer than 6 years [8, 9]. The reason for this difference is unknown [9]. Median survival is believed to be 3-5 years [94]. There is currently no therapy that reverses or cures the lung damage [93, 96]. Lung transplantation remains the only available treatment if the patient meets established criteria [7, 93].

Patients diagnosed with IPF share problems common to those with a chronic illness. Chronic illness, a permanently altered health state, results from a non-reversible pathological condition [11]. The consequence is a residual disability that cannot be corrected by a simple surgical procedure or cured by medical therapy [12]. The diagnosis of a chronic illness can

affect the emotional response, sometimes in a manner disproportionate to the extent of physical disability. This can be a major life change for the patient and family [13]. Consequently, it is essential to treat psychosocial as well as physical symptoms.

As with other chronic illnesses, the progressive disability associated with IPF impacts both the patient and care partner. The impact can be particularly devastating given the rapidly progressive nature of the decline, variability across individuals, and lack of effective therapy. DeVries and colleagues [14] assessed perceptions of health-related quality of life (HRQoL) in 41 patients with IPF and the relationship between HRQoL, depressive symptoms, and breathlessness. They reported that the HRQoL was mainly impaired in the domains of "physical health" and "level of independence" compared to matched healthy controls. Of note, approximately 25% of patients with IPF indicated that they experienced depressive symptoms.

These findings and other reports describing response to this diagnosis[51-53, 55, 56, 97] suggest that patients with IPF may benefit from an intervention that provides assistance coping with the consequences of this diagnosis [15]. Palliative care should be offered from the time of diagnosis, even while the patient pursues treatments to slow or stabilize the disease [7]. Although palliative care is commonly integrated into the management of patients with cancer, its use in patients with advanced lung disease is more limited. Nevertheless, palliation may be the only therapeutic option for many diseases of the lung, including lung cancer, chronic obstructive pulmonary disease, cystic fibrosis and IPF [17]. The concepts integral to palliative care seem particularly appropriate for patients with IPF who face a mortality rate that is greater than many cancers [98].

No studies were identified that tested the ability of a brief intervention incorporating palliative care concepts to assist patients with IPF to deal more effectively with their illness.

Accordingly, we developed a 6-session *disease management intervention* (PRISIM: Program to Reduce Symptoms and Improve Lifestyle Management) designed to be delivered to the patient and care partner. Because chronic illness impacts the patient and care partner, both individuals were included as participants in all sessions of the program.

The purpose of this study was to test the ability of PRISIM to decrease symptom burden, decrease perceived stress, and improve perceptions of HRQoL for patients with IPF and their care partners. The aims were to: 1) determine the short term (6-week) impact of a disease management intervention on symptom burden (shortness of breath, anxiety, depression), perceived stress, and HRQoL in patients with IPF; 2) determine if participation in a disease management intervention altered caregiver burden as evidenced by changes in anxiety and depression, perceived stress, and HRQoL; and 3) evaluate the common themes of the experience of living with IPF from the perspective of the patient and care partner. We hypothesized that participation would improve perceptions of HRQoL and decrease symptom burden.

5.3 METHODS

5.3.1 Design

The study enrolled 42 participants (Figure 5). Ten patient/care partner dyads were randomized to the intervention group and 11 patient/care partner dyads to the usual care group. Both groups completed questionnaires designed to measure anxiety, depression, perceived stress, and HRQoL at two intervals: study entry (baseline) and 7 weeks later. Patients in both groups completed a questionnaire to assess dyspnea at the same intervals. To provide additional insight into the

experience of participation in the intervention, patient/care partner dyads randomized to the intervention were interviewed in their homes, using a qualitative approach, 3-9 months after study completion. The study was reviewed by the University Institutional Review Board and all participants provided informed consent.

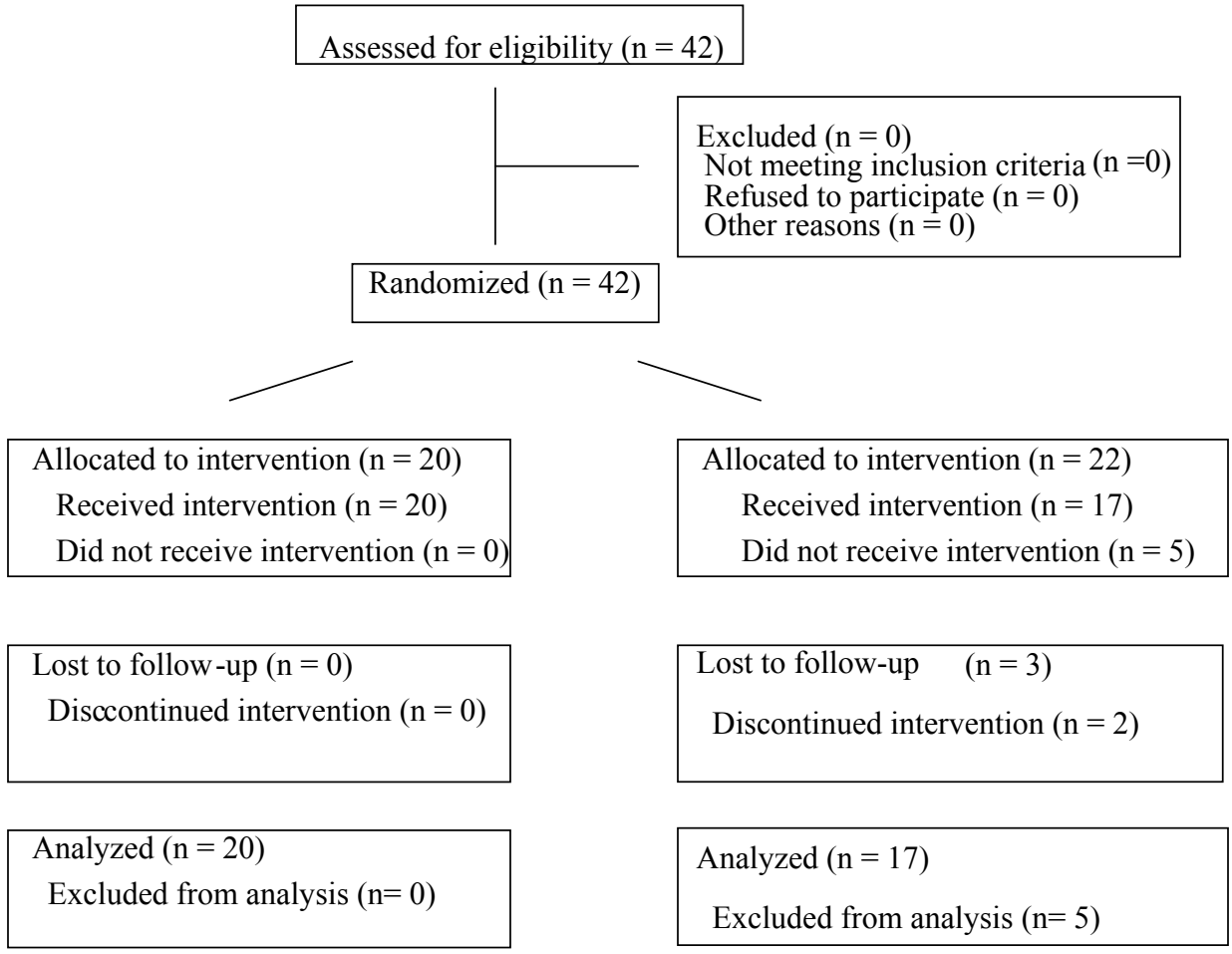

Figure 5. Recruitment Criteria

5.3.2 Setting & Sample

Patients were recruited from the Simmons Center for Interstitial Lung Disease located within the University of Pittsburgh Medical Center. To be eligible, patients were required to be: 1) 21 years or older; 2) able to read and understand English; 3) diagnosed with IPF; and 4) have a forced vital capacity (FVC) consistent with moderate (FVC 55-70% predicted) or severe (FVC < 55% predicted) disease. Care partners were required to be: 1) 21 years or older; 2) able to read and understand English; and 3) live with and/or care for the patient with IPF. To enroll, both the patient and care partner had to consent to study participation. Group entry was determined using randomly numbered cards that were placed in sequentially numbered opaque envelopes and opened after consent was obtained and baseline questionnaires completed.

5.3.3 Intervention

The intervention consisted of 6 weekly group sessions attended by patients and care partners. Each session was two hours in length and took place in a conference room located in the health care facility. The sessions were led by a group leader trained as a pulmonary clinical nurse specialist (Sessions 1, 5, & 6), psychiatric clinical nurse specialist with training as a cognitive behavioral therapist (Session 2 & 3) and advanced care planning instructor (Session 4).

The sessions included: Session 1: "What is IPF and How to Live with it?" reviewed the causes, pathophysiology, and treatment of IPF. Participants received a copy of the book, "Feeling Good" that was used for assignments and read by the patient or care partner at their leisure [88]. Session 2: "Gaining Control of Your Moods and Feelings: You Feel the Way You Think" began with a discussion of the basic principles of cognitive behavior techniques including the negative triad

and the role of automatic thoughts. Ten cognitive distortions were discussed and group members were prompted to share examples of how they used distortions in their own lives. Group members were encouraged to discuss these distortions with other group members by giving concrete examples related to their lung disease. Short homework activities were assigned. Session 3: "Gaining Control of Your Moods and Feelings: What can you do about Depression" began with the group leader asking members to share examples from their homework. Members were encouraged to provide feedback to the other group members. The group leader discussed the concepts of stress and depression and interrelationships with illness for patients and care partners. Treatment modalities for depression and benefits gained from effectively dealing with depression were discussed. Session 4: "Putting Your Life in Order: What Do I Do Now?" began with group members being asked to share their observations about life and planning for uncertainty while trying to maintain control. The group leader addressed important end-of-life issues including struggling with the reality of terminal illness, working with the medical team, helping family and loved ones cope with the illness, and planning ahead to set their affairs in order. Session 5: "Living with IPF" focused on living with IPF. Using lecture and group discussion, symptom management, energy conservation, correct use of oxygen therapy, and the importance of exercise were addressed. Session 6: "Wrap-up and Review Group" encouraged group members to share what they learned during past sessions and how they could apply this to their lives. Control group participants usual care and a copy of the book given to experimental group participants. [88]

5.3.4 Instruments

Dyspnea was measured using the *University of California at San Diego Shortness of Breath Questionnaire (SOBQ)*. [21] The SOBQ is completed by asking subjects to rate the severity of shortness of breath on a 6-point scale (*0=not at all to 5=maximal or unable to do because of breathlessness*) during 21 activities of daily living associated with varying levels of exertion. There were 3 additional questions that asked about daily life limitations due to shortness of breath, fear of over exertion and fear of shortness of breath. If subjects did not routinely perform the activity, they were asked to estimate the shortness of breath anticipated. The score was obtained by summing responses on the 24 items to form a total score (range 0-120). Internal consistency and validity have been well established [21].

Anxiety was measured using *The Beck Anxiety Inventory (BAI)*, a standardized, 21-item tool designed to assess the presence and severity of common symptoms of anxiety, both psychological (e.g., feeling nervous, scared) and physical (e.g., heart pounding, hands trembling) [76]. The tool uses a 4-point scale (0 = absent/not at all disturbing to 3 = I could barely stand it). Items are summed and total scores range from 0 to 63. A scores of 0 to 7 indicates no anxiety, 8 to 15 mild anxiety, 16 to 25 moderate anxiety, and scores 26 or higher, severe anxiety [76]. Reported mean BAI scores for psychiatric outpatients with Diagnostic and Statistical Manual of Mental Disorders, Revised Third Edition diagnoses of anxiety disorders range from 17 to 29 [77]. Means for community samples have ranged from 7 to 11.

Depressive symptoms were measured using the *Beck Depression Inventory-II (BDI* [80]*)*. The BDI-II, a 21-item self-report instrument, is designed to measure the severity of depression in adults and adolescents 13 years or older [80]. The BDI-II is a revised version of the original

instrument. A score of 0-13 suggests minimal depression, 14-19 mild depression, 20-28 moderate depression and 29-63 severe depression [81].

The *Perceived Stress Scale (PSS)* [72] was designed to measure the degree to which subjects find their lives unpredictable, uncontrollable, and overloading. Individuals are asked to indicate how they feel or thought in a certain way in the last month using the 5 options, 0 (never) to 4 (very often). The range of scores is from 0 to 40, with higher scores indicating more appraised stress. There are no cut-off points on the PSS. The internal consistency and validity have been well established with a diverse population [83-85].

HRQoL was measured using the *SF-36* (Short Form Medical Outcomes Study Form) Version 2, a widely used generic scale, that has been used to assess HRQoL in a variety of chronic medical conditions and validated in patients with IPF [47, 52] . The SF-36 assesses 8 domains of HRQoL. Scores range from 0 = maximum impairment to 100 = no impairment. Scores on the 8 domains can be grouped into a physical component score (PCS) that includes scores for physical functioning, role physical, bodily pain, and general health and a mental component score (MCS) that includes scores for vitality, social functioning, role emotional, and mental health [86]. The PCS and MCS was normalized to responses from the general population (mean score is 50). The validity, reproducibility, and responsiveness of the SF-36 have been well-demonstrated [87]. The total time required to complete the questionnaire battery was approximately one hour. Demographic & Medical Profile data obtained from the patient and care partner included age, gender, race, and education. Diagnosis and pulmonary function values (FVC) were obtained from the medical record.

5.3.5 Interviews

Interviews of the dyads who participated in the intervention were conducted in their home 3-9 months after the intervention by a member of the research team (KOL). These interviews were open-ended, which allowed for data collection that reflected the perspectives of each member of the dyad regarding their individual experience of participating in the intervention. Responses guided future questions and became more focused as the interviews progressed.

5.4 DATA ANALYSIS

5.4.1 Quantitative analysis

Baseline demographic and medical profile data were compared using Chi-square and t-tests, as appropriate. Because differences in intervention and control groups were found at baseline, questionnaire responses were analyzed using analysis of covariance (ANCOVA). SPSS v14.0 statistical software was used for all data analyses. A p-value of less than 0.05 was chosen to indicate statistical significance.

5.4.2 Qualitative analysis

Sample size was determined by the number of interviews necessary to achieve theoretical saturation, where the same information continues to be validated and less and less new information is elicited. The sample included 8 patient/care partner dyads. One care partner was

interviewed alone as the patient died prior to scheduling the interview. Verbatim transcripts were analyzed line by line initially and then more generally by comparing transcripts to transcripts guided by a qualitative research expert (EO). Data analysis consisted of open coding, wherein codes or concepts were developed that reflected the meaning in the transcripts. During open coding the researchers were very inductive and open to any and all codes that possibly reflected the meaning in the data. These codes were then analyzed through a process referred to as axial coding, in which the complexities of each code were explored, e.g., conditions under which the codes occurred and consequences of these codes. Eventually, only certain codes were supported in the ongoing data analysis and those codes were then analyzed in more depth through a process called selective coding. Those selective codes were then integrated into an explanatory framework to reflect the overall meaning in the data.

5.5 RESULTS

Of the 42 subjects, two patients and three care partners did not complete data collection; all were enrolled in the control group. One patient died during the period between pre and post intervention, and the second patient did not return to complete the questionnaires. Their care partners and one additional care partner did not return to complete the questionnaires. Study data are reported for the remaining 37 participants. No significant differences were found between the intervention and control group in regard to age, gender, race, education (Table 6). Slightly more than half (58%) of patients had scores indicating mild to severe anxiety compared to 21% of care partners. Two patients (10.5%) reported scores consistent with mild depression and two scores consistent with moderate depression. No care partners reported depressive symptoms.

Table 6. Demographic characteristics and medical profile of patients with IPF and their care partners who provided pre and post intervention data (n=37)*

	Patients		Care Partners	
	Intervention (n=10)	Control (n=9)	Intervention (n=10)	Control (n=8)
Age, years	65.2 ± 10.3	67.1 ± 11.9	63.3 ± 12.1	67 ± 9.1
Gender, % male	76.2%	54.6%	23.8%	45.5%
Race, % Caucasian	100%	91%	100%	91%
Education, years	13.9 ± 2.6	14.5 ± 2.9	14.2 ± 2.0	14 ± 2.1
FVC % predicted	61.8 ± 10.8	67.6 ± 23.1	N/A	N/A
Method of Diagnosis Biopsy, % CT Scan, %	 70% 30%	 82% 18%	N/A	N/A

* Values are mean ± SD unless indicated

5.5.1 Patient response

When post intervention comparisons were made using ANCOVA, controlling for pre intervention scores, there were no significant differences for mean scores on the SOBQ (p=.972), PSS (p=.531), BDI (p=.894), or SF-36 Mental Component (p=.772) (Table 7).

Table 7. Adjusted Mean Scores reflecting symptom burden, stress and perceptions of HRQoL of intervention and control group participants (n=37)*

	Patients		Care Partners	
	Intervention	Control	Intervention	Control
SOBQ	49.51 ± 22.64	49.88 ± 22.64		
Beck Anxiety	15.13 ± 6.92	8.56 ± 6.95	2.22 ± 4.28	4.72 ± 4.32
Beck Depression	9.71 ± 4.34	9.44 ± 4.35	4.33 ± 3.29	4.84 ± 3.29
Perceived Stress	19.32 ± 3.64	18.20 ± 3.65	17.61 ± 2.68	20.99 ± 2.69
SF 36 Physical	31.06 ± 4.61	36.04 ± 4.63	50.12 ± 5.72	51.31 ± 5.72
SF 36 Mental	55.98 ± 2.71	55.61± 2.71	50.52 ± 1.67	50.05 ± 1.67

*Values are mean ± standard deviation.
Definition of abbreviations: SOBQ = shortness of breath questionnaire

Mean differences of BAI scores approached statistical significance (p=.077) (Figure 6).

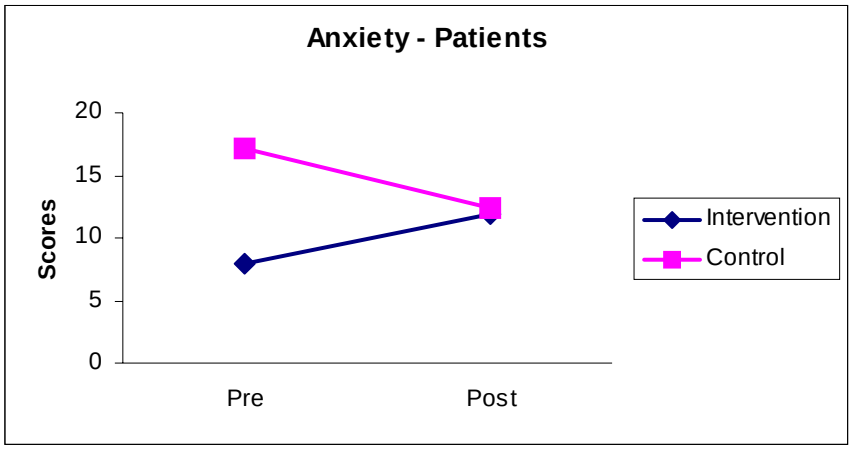

Figure 6. Change in anxiety scores for experimental and control group patients prior to and following the intervention.

The adjusted mean BAI score for the intervention group was nearly 6.5 points higher than the control group. Mean scores for the SF-36 physical component exhibited a statistically significant difference (p=.038) (Figure 7); mean scores were approximately 5 points lower for intervention patients.

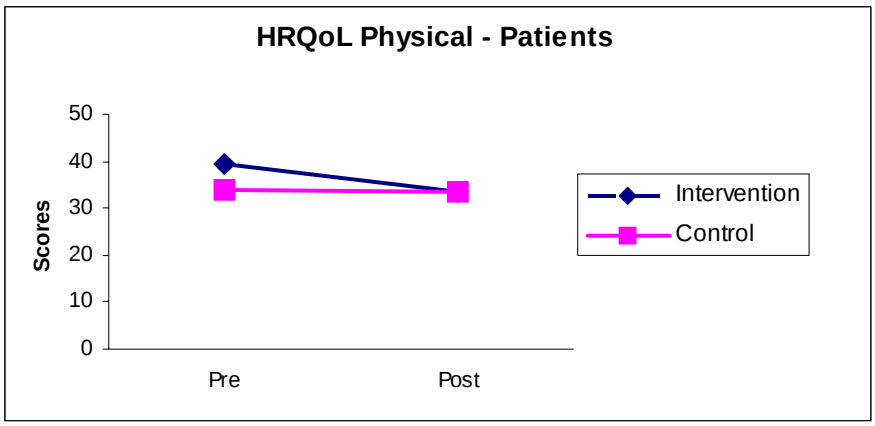

Figure 7. Change in HRQoL physical component scores for experimental and control group patients prior to and following the intervention.

5.5.2 Caregiver response

When post intervention comparisons were made using ANCOVA, there were no significant differences between intervention and control group caregivers for mean scores on the BAI (p=.260), BDI (p=.751), SF-36 physical component (p=.669), or SF-36 Mental Component (p=.565). Mean scores on the PSS were significantly lower (p=.018) for the intervention group (Figure 8). Accordingly, findings suggested no impact on anxiety, depressive symptoms, or HRQoL, but a positive impact on perceived stress for care partners in the intervention group.

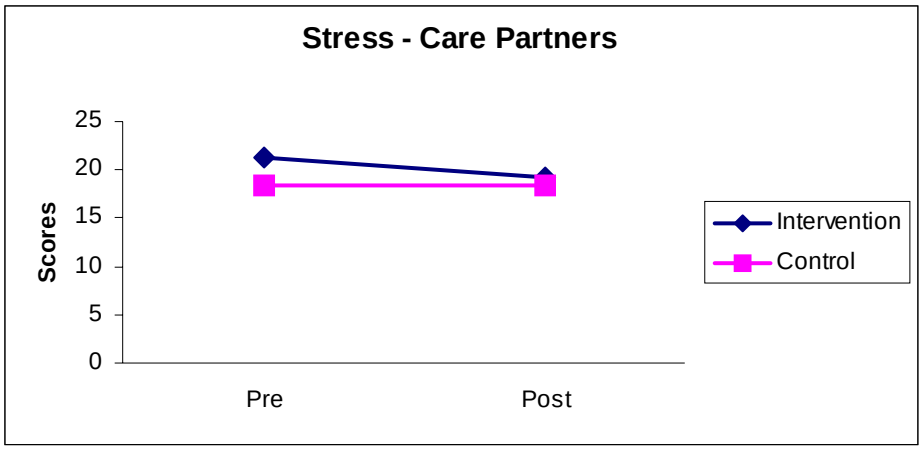

Figure 8. Change in stress scores for experimental and control group care partners prior to and following the intervention.

5.6 QUALITATIVE ANALYSIS

Three common themes emerged after analyzing the verbatim transcripts. These themes included "didn't feel isolated when participating in a disease management program", "able to put my disease into perspective", and "felt it was important to participate in research to help others with the disease". The participants shared that they recognized "you aren't the only one that has a problem" which "gave comfort" and provided an "improved mental picture". The opportunity to be with others with the same diagnosis was viewed as "very beneficial" and led to perceptions that one was "not as bad off as you think you are". There was also a sense that participation "might help us and maybe we could help somebody else". These perceptions are reflected in the following statement of a patient participant: "First and foremost, it's always good to be around people that have the same illness who share the same concerns, your same fears, your same outlook, what is to be perceived in the future for yourself, everybody comes from different

aspects of life, different ages, and the opportunity is there to express your innermost feelings or figure out how you can address your issues in life".

5.7 DISCUSSION

To our knowledge, this study is the first to test ability of an intervention to decrease symptom burden and improve perceptions of HRQoL for patients with IPF and their care partners. Our findings were not anticipated. Ratings of physical HRQoL were lower (more negative perception) compared to controls at the conclusion of the intervention. After controlling for baseline anxiety level, patients in the intervention group also tended to exhibit higher scores (more anxiety) than the control group. As anticipated, care partners in the intervention group experienced less stress following participation.

We were challenged in interpreting these findings. Although the outcome suggested increased distress, comments were uniformly positive. Patients and care partners in the intervention group repeatedly voiced appreciation for being able to participate in the PRISIM program. Our qualitative findings supported these statements. The common themes that emerged endorsed feelings of less isolation, a more balanced view of the illness, and personal satisfaction from being able to participate in research to help others. Using a mixed method quantitative and qualitative approach allowed us to capture the complexity in the data; that is, although there was some increase in distress with the PRISIM program, there were simultaneously, benefits that were acknowledged by the participants who took part in the PRISIM program.

This outcome is perhaps not surprising. The PRISIM program was constructed to enhance knowledge of the disease and symptom management and assist patients to deal

constructively with depressive symptoms and address beliefs in regard to end-of-life decision-making. While it is possible to avoid such discussions, the rapid progression of IPF suggests that a more proactive approach is necessary. Of note, between the interval when the intervention was completed and qualitative interviews were scheduled, 6 patients died and 7 received a lung transplant. When patients are faced with a terminal illness or a life changing event such as a lung transplant, frank disclosure can open the door to discussion regarding palliative and terminal care preferences [99]. Unfortunately, such discussions are often avoided [100, 101]. From a study of 105 patients with chronic obstructive pulmonary disease, Heffner and colleagues [101] reported that most patients (94.3%) expressed worries about their health although few (42%) had completed an advance directive. Although most (98.9%) wanted discussions about end-of-life decision-making, few (19%) had such discussions and few (14.3%) thought their physician understood their preferences. As a consequence, patients were left to wonder what they might experience and clinicians were left without clear knowledge of their preferences. Not discussing such issues can lead to distress on the part of the patient and care partner that may not be recognized by clinicians. Findings of this study reinforce the importance of directly addressing such topics and carefully listening to the patient and care partner to elicit what it is they believe is beneficial.

Few studies have addressed how to assist patients to cope with the depressive symptoms that commonly accompany chronic illness. Hopwood et al. [33] studied psychological distress in patients with advanced lung cancer and reported a prevalence of the affective disorder varying from 23% - 47%. Depression was found in patients with more advanced stages of malignancy, more severe illness, and poorer physical status. This mood disorder may be in part a reaction to the news of the diagnosis, but it may persist, causing an added burden during any treatment, and

may interfere with symptom control, compliance with treatment, and possibly reduce survival [33]. Patients with lung cancer are similar to patients with IPF in regard to their prognosis and limited life expectancy. In 41 patients with IPF, DeVries and colleagues [14] reported that approximately 25% experienced depressive symptoms. Our sample reported a lower incidence. Potentially, this finding was influenced by the support available at our Center, including a Pulmonary Clinical Nurse Specialist, support group, and active research program.

Cognitive behavioral techniques have been widely used to managed depressive symptoms following a diagnosis of Alzheimer's [102, 103], end-stage lung disease [103] , arthritis [104], coronary artery disease [103], and in older adults (10). Cognitive behavioral interventions can benefit those who suffer from major depression, as well as those who experience depressive symptoms and anxiety [47, 48]. Patients with recurrent depression who were maintained at the same dose of their antidepressant medication, but who received six cognitive therapy sessions, had a lower relapse rate than those who only had their antidepressant medication dose increased and did not receive cognitive therapy [48]. The Program to Encourage Active, Rewarding Lives of Seniors (PEARLS) study, which tested a cognitive behavioral intervention in patients with minor depression, found that patients who enrolled in the program had a 50% reduction in depressive symptoms, 43% had complete remission from depression, and 36% had improved HRQoL [105]. No studies were identified that studied the impact of cognitive behavioral techniques in patients with IPF.

5.8 LIMITATIONS

The sample size was small and findings need to be confirmed in a larger sample followed for a longer time. The requirement to attend a 2-hour session for 6 consecutive weeks, in addition to two additional sessions to complete instruments, was a barrier to recruitment. An internet based intervention would likely be more appealing. However, the selected format had a major advantage in promoting discussion with other participants and nurse group leaders. Of note, some of the participants traveled 2-3 hrs to attend the session, thus they had to plan ahead to make sure they had adequate oxygen. The rapid progression of this disease also served as a limitation. At 9 months after study entry, 7 patients received lung transplants, 6 died, and 3 were awaiting lung transplantation. This outcome occurred despite the fact that all participants had a similar decrement in lung function at entry into the study (FVC 64.7 ± 17.79). The great variability in disease progression reinforces the need to develop interventions that prepare patients for end-of-life decision-making and dealing with depressive symptoms that often accompany chronic illness. .

5.9 CONCLUSION

The PRISIM program is innovative in its attempt to combine information about disease management with information commonly included in palliative care programs and strategies designed to elicit discussion of preferences in end-of-life decision-making. The program enrolled the care partner and patient in an attempt to enhance sharing of knowledge and beliefs on topics that might be difficult to discuss in other settings. The use of palliative interventions and advance

care planning can assist patients with IPF and their care partners to more successfully cope with the consequences of this disease. Further study is needed to further evaluate the positive and negative outcomes of this intervention.

APPENDIX A

PRISIM

PRISIM

A Program to **Reduce**
Idiopathic Pulmonary Fibrosis
(IPF) Symptoms
and

Improve Management

Dorothy P. & Richard P. Simmons
Center for Interstitial Lung Disease

University of Pittsburgh

2004

PRISIM

Session I
"What is IPF and How to Live with it?"

OUTLINE

I. What is IPF?
II. How is it diagnosed?
III. Why did I get it?
IV. Who else gets it?
V. How is it treated?
VI. How do I live with the disease?

Content:

1. How your lungs work.
2. What happens you have IPF.
3. Dealing with cough and shortness of breath
4. Using prescribed oxygen and other medicines.
5. Using oxygen safely
6. Importance of Exercise and good nutrition.
7. Smoking Cessation
8. Avoid irritants
9. Energy conservation
10. Preventing lung infections
11. Stress and Relaxation
12. Dealing with feelings
13. Getting the support that you need.

Handouts:

"What is Idiopathic Pulmonary Fibrosis"
"Understanding Interstitial Lung Disease"

PRISIM

Session II

Part I

Gaining Control of Your Moods and Feelings

"You Feel the Way You Think"

OUTLINE

I. Negative moods and feelings
II. Cognitive distortions (Twisting your thoughts)
    Automatic Thoughts (Self-criticism)
    vs. Rational Responses (Self-defense)
III. Ten things you should know about your anger (HO)
IV. Your IQ (Irritability Quotient)
V. Ways of defeating guilt
VI. Sadness is not depression
VII. How do you know if you're depressed?

Homework:
Dysfunctional Attitude Scale
Daily record of dysfunctional thoughts and attitudes
Pleasure predicting

PRISIM

Session III

Part II

Gaining Control over your Moods and Feelings

OUTLINE

"What can you do about Depression."

I. Homework review
II. Motivation: Practical "little steps"
III. What you need to know about treatments

   a. Cognitive Behavioral Therapy
   b. Relaxation Techniques
      1. Guided Imagery
      2. Music
      3. Exercise
   c. Antidepressants (12 myths and what you need to know)

IV. The Ultimate Victory: Putting it all together and Choosing to live.

PRISIM

Session IV

Putting Your Life in Order

OUTLINE
"What do I do now?"

I. *An unexpected variety of new possibilities*

A. Observations about life: Searching for meaning
B. Planning for uncertainty while trying to maintain control

1. Taking care of yourself
2. Setting realistic goals
3. Issues to keep in mind
4. Struggling with the reality of your illness:
   An emotional roller coaster ride (Kubler-Ross)

II. Working with your "team" while dealing with the medical bureaucracy

A. Getting the help you need
B. Talking with your doctor

III. Helping your family and loved ones
IV. Things to do when time may be short
V. Setting your affairs in order: Planning ahead

A. Personal activities
B. Legal issues

VI. Quality of life for the spiritual traveler

Handouts:

UPMC Advanced Directives

PRISIM

Session V

Living with IPF

OUTLINE

I. Symptom Management
   1. Dealing with Cough
   2. Managing Shortness of Breath

II. Energy Conservation

"Saving your energy for what you like best"

III. Correct Use of Oxygen Therapy

   1. Interpreting your study results
   2. Following the doctor's prescription
   3. Types of Equipment

IV. Importance of Exercise

   1. "Yes, Exercise is Possible"

Handout:

"How to use Oxygen Therapy"

PRISIM

Session VI

OUTLINE

Wrap – Up

I. Review of Key Concepts

II. Questions/Discussion

III. Questionnaires

APPENDIX B

PRISIM INTERVIEW

PRISIM Interview

1. Tell me what it was like for you to come to this course (pt) (care partner).

   What was the experience like of being a part of this course?
   How would you describe this course?

2. What kinds of things did you anticipate in choosing to participate(pt) (care partner)?

   What kinds of things, if any, did you anticipate to be difficult for you in participating in this course?
   What kinds of things, if any, did you anticipate to be helpful for you in participating in this course?

3. Tell me what your experience was in participating in each of the series:

   - "How to Live with the Disease"
   - "Dealing with Feelings"
   - "Advanced Planning"
   - Wrap-up
   - Nocturnal Oxygen Saturation Study (NOSS)

4. Did you make any changes in your management of your illness as a result of the findings from your NOSS? If yes, please describe any changes you experienced?

5. Overall, how did this course have an impact on your life, if it did.

6. Please add anything else that I have not asked that you believe is relevant to our interview.

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