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INTERSTITIAL LUNG DISEASE (A. HAJARI CASE, SECTION EDITOR)

Nonpharmacological Therapies for Interstitial Lung Disease

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Abstract

Purpose of Review Interstitial lung diseases (ILDs) cause unpredictable degrees of fibrosis and inflammation in the lungs leading to functional decline and varying symptom burden for patients. Some patients may live for years and be responsive to therapy, and other disease trajectory may be shorter and similar to patients with lung cancer. This ultimately affects the patient's quality of life as well as their caregiver(s).

Recent Findings Nonpharmacological therapies play an important role in treatment of interstitial lung disease. These include symptom management, pulmonary rehabilitation, oxygen therapy, and palliative care. While ILDs are associated with high morbidity and mortality, different models of care exist globally. New tools help clinicians identify and address palliative care needs in daily practice, and specialty nurses and ILD centers can optimize care.

Summary This paper provides an overview of nonpharmacological therapies available for patients with interstitial lung disease.

Keywords Interstitial lung disease · Nonpharmacological therapy · Symptom management · Pulmonary rehabilitation · Oxygen therapy · Palliative care

Introduction

Interstitial Lung Disease

Interstitial lung diseases cause varying degrees of fibrosis and inflammation in the lungs of patients [1], and the disease progression leads the patient to experience functional decline and varying symptom burden, ultimately affecting quality of life. It is critical that the clinician is able to make a confident diagnosis of the specific form of ILD and formulate a patient-centered, personalized management plan to achieve remission or stabilization of the disease process when possible [2].

It is important for providers to relay disease information in a timely manner that is easily understood by patients and their caregivers to help them best manage their ILD. The words used to describe the major scarring lung diseases include interstitial lung disease, idiopathic pulmonary fibrosis (IPF), hypersensitivity pneumonitis (HP), etc.; these terms confuse not only patients but also other medical providers [3].

Patients with interstitial lung disease experience a wide range of diagnoses and can benefit most from early evaluation at a center with ILD expertise, such as the Pulmonary Fibrosis Foundation (PFF) Care Center Network [4]. Connecting patients with access to clinical trials and support groups can provide additional benefit to participate in research studies to advance knowledge and treatment of ILD's, provide accurate information about their disease, and help to connect with other patients with similar needs. These measures can help to improve social support and help the patient and caregiver avoid social isolation ultimately improving their quality of life. Early educational programs can help to increase knowledge of the disease so that patients and their caregivers can have a better understanding of the effect and consequences of these relentlessly progressive diseases [3, 5•]. Participation in support groups offers the patients and their caregiver(s) the opportunity to receive additional education and support outside of the office visit. Support groups offer resources to teach individuals how to cope and adapt to the lifestyle that is often dictated by their illness. The Pulmonary Fibrosis Foundation provides a list of support groups that are local and international, and on-line and telephone-based communities [6].

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Nonpharmacological therapies play an important role in the treatment plan for patients diagnosed with interstitial lung disease (ILD).

Symptom Management

Patients with ILD have a wide range of diagnoses and prognoses; some may live many years with a disease that is responsive to treatment, but in those patients with progressive idiopathic fibrotic interstitial lung disease (PIF-ILD), the disease trajectory is shorter and similar to that of lung cancer [7••, 8]. Despite the varied nature of ILD, patients experience common symptoms related to their chronic lung disease which contribute significantly to their morbidity and impact their quality of life [9]. In a quantitative review of patients with PIF-ILD, the overwhelming majority of patients had breathlessness (68.2–98%), cough (59–94%), heartburn (25–65%), and depression (10–49%) [7••]. In addition, this review found that patients experience a wide array of constitutional symptoms including sleep disturbances, fatigue and weight loss, and anorexia [7••]. The psychological stress of having a chronic life-limiting illness can complicate symptom control requiring effective symptom management, best achieved by a multidisciplinary approach that incorporates patient education and self-management to articulate goals of care and treatment plans [9]. A number of both pharmacologic and nonpharmacologic therapies are available to reduce symptoms in patients with IPF. These include low-dose narcotics, pulmonary rehabilitation (including pursed lip breathing) for treatment of dyspnea, and supplemental oxygen [10, 11]. Dyspnea and cough often improve with supplemental oxygen. Cough is challenging to treat, but distressing to patients and caregivers. Treatment options include a range from hot tea, honey, menthol lozenges to treatment of gastroesophageal reflux, postnasal drip medication, benzonatate, and opiate-containing medications [12]. Early identification of symptoms and referral for palliative care to alleviate symptom burden and improve quality of life are crucially important treatment goals [13]. Pulmonary rehabilitation also plays an important role in symptom management [9].

Pulmonary Rehabilitation

Pulmonary rehabilitation (PR) has demonstrated physiological, symptom reducing, psychosocial, and health economic benefits for patients with chronic respiratory disease [14]. Patients with ILD experience reduced functional capacity, dyspnea, and exercise-induced hypoxia [15]. Referral to PR includes exercise training that has shown improvement in long-term outcomes in ILD including 6-min walk distance (6MWD), dyspnea, health-related quality of life, and peak exercise capacity for patients with ILD [16, 17]. In a study of 142 participants with different ILD's (IPF, asbestosis, connective tissue disease-related ILD, and other causes of ILD), participants were randomized to 8 weeks of supervised exercise training or usual care. Those participants who participated in supervised exercise training significantly increased their 6MWD and health-related quality of life, with more lasting effects in those with milder disease [16]. In another study done at 3 PR centers in North America, PR improved functional capacity and quality of life in patients with a variety of ILDs, with benefits lasting for at least 6 months [17]. This group reported that the "consistency and magnitude of benefit across endpoints is substantial and markedly better than pharmacological interventions that have been studied in these diseases" and suggest that pulmonary rehabilitation should be the first line of therapy for patients with ILD [17]. Barriers for participation in PR may include distance from patient's location and reimbursement for attendance. Reimbursement may involve getting authorization from the patient's payor source. Patients may qualify for cardiac rehabilitation; while the focus may differ between cardiac and pulmonary, the emphasis is on supervised, safe exercise.

In another study of patients with ILD attending pulmonary rehabilitation, patients wanted ILD-specific content and wanted information about end-of-life planning and most were happy to discuss it in a group. In that same study, clinicians supported discussion of advanced care planning but not necessarily in the pulmonary rehabilitation setting [18]. Communication with patients about goals of care is crucial, and continued research is needed in this area.

Oxygen Therapy

In the 1980s, use of supplemental oxygen therapy increased after the NOTT (Nocturnal Oxygen Therapy Trial) and the MRC (Medical Research Council) trials demonstrated survival benefits of providing long-term oxygen therapy to patients with resting arterial partial pressure of oxygen consistently less than 55 mmHg [19, 20]. Today, more than one million people in the USA use long-term oxygen therapy, the majority with chronic obstructive lung disease (COPD) [21]. Oxygen therapy is the most frequently used treatment for patients with ILD and IPF to treat hypoxemia and halt progression or prevent development of hypoxia-induced pulmonary hypertension, cardiovascular morbidity, or cognitive dysfunction [22, 23].

Oxygen prescriptions vary greatly for patients with ILD with patients often requiring oxygen with exertion and sleep earlier in the disease process before they require oxygen with rest. In one study, exertional hypoxemia was found to be more severe for patients with fibrotic lung disease than those patients with COPD. This group compared results of a 6-min walk test (6 MW) performed on room air in 134 patients with ILD and 247 patients with COPD. Diffusing capacity (DLco) was the strongest predictor of desaturation in both cohorts

with ILD patients experiencing greater oxygen desaturation during the 6 MW compared to patients with COPD [24•].

Supplemental oxygen has allowed patients who otherwise would be homebound to be more mobile, work, exercise or attend pulmonary rehabilitation, travel, care for family members, and also experience improvement in their symptoms, including dyspnea, ultimately improving their quality of life [25, 26]. Patients who require supplemental oxygen, especially those with ILD, experience frequent and varied problems with receiving adequate portable systems to meet their dose requirement. Oxygen equipment is bulky and options are limited as oxygen dose increases. In a study of 30 clinically stable ILD patients with varying disease severity, carrying portable oxygen versus using oxygen from a stationary concentrator resulted in significantly greater dyspnea and shorter distances in timed testing [23]. Portable systems can deliver continuous flow (CF) and intermittent flow (IF), and while IF devices are safe and generally effective in correcting hypoxemia, there is variability in delivery and patient response, and therefore, patients need to be tested on these devices [26]. Use of a pulse oximeter is recommended to allow patients to adjust their oxygen flow to maintain saturations > 89% at all times [27].

In a recent ATS survey of 1926 patients with lung disease who used oxygen, individuals with IPF were rarely on oxygen more than 5 years, but frequently used oxygen > 5LPM [28•]. Patients who received oxygen education had less health care utilization, including emergency visits and hospitalizations [28•]. Use of "a detailed discussion that includes an educational overview of oxygen, recommendations for how to use oxygen correctly, and disclosure of what hardships and benefits the patient (and their caregiver) might expect from oxygen" is endorsed for patients prescribed oxygen therapy [27]. The process of oxygen prescription can be improved by providing patients with clearer expectations and trustworthy educational resources [29].

Palliative Care

ILDs are highly disabling and patients experience a loss of functional ability, resulting in great symptom burden as the disease advances ultimately impacting the patient's quality of life [11]. Patients with ILD often suffer unmet physical and psychological needs as they live with their ILD [30].

Palliative care is the comprehensive treatment of the discomfort, symptoms, and stress of serious illness and should be started as soon as the patient is diagnosed. Symptom management is relevant even in patients with mild to moderate disease. Traditionally, palliative care was seen as replacing the curative care with palliative or end-of-life care, but the paradigm has shifted and palliative care should be offered alongside all other treatments for the disease [9, 31]. See Fig. 1. In prior studies, patients with advanced lung disease, including IPF, have been found less likely to receive palliative care than malignant diseases and other chronic conditions, including dementia [32, 33]. Several reasons for fewer referrals include uncertainty regarding prognosis, lack of provider skill to engage in discussions about PC, fear of using opioids among patients with chronic lung disease, fear of diminishing hope, and perceived and implicit bias against patients with smoking-related lung disease [34].

The main goal of palliative care (PC) for patients with interstitial lung disease (ILD) is to improve and maintain quality of life. Optimal quality of life care includes symptom-centered approaches, best supportive care, caregiver-centered

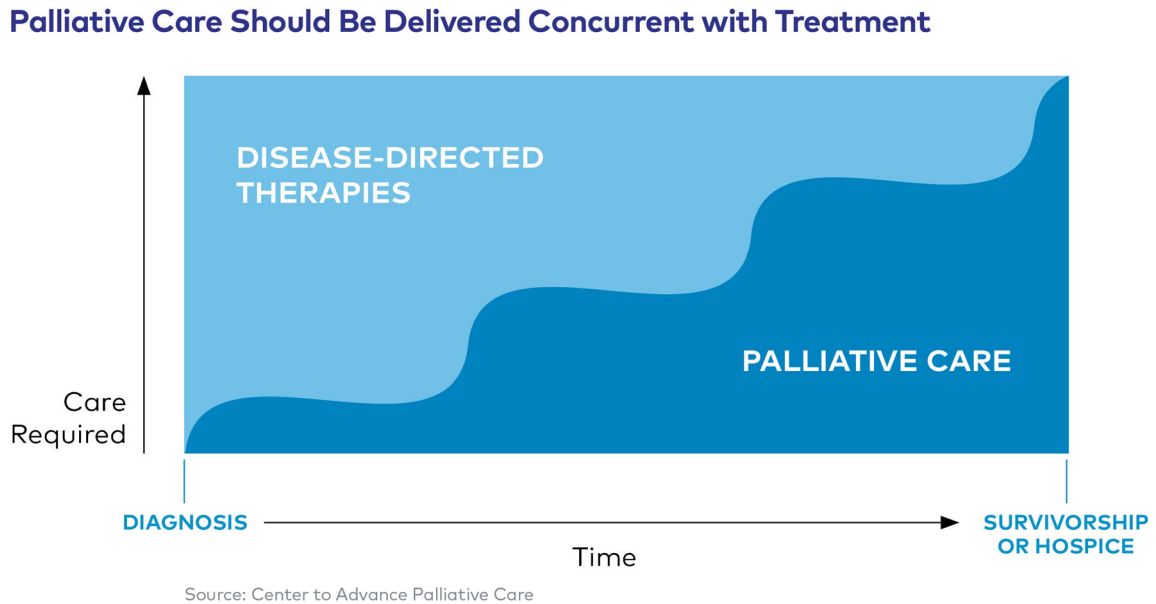

Fig. 1 Palliative care's place in serious illness. Used with permission from Center to Advance Palliative Care

Table 1 The needs assessment tool: progressive disease in interstitial lung disease (NA:PD-ILD). Copyright Hull York Medical School, University of Hull, UK

Needs Assessment Tool: Interstitial Lung Disease

Name:

(or attach address label)

Please complete all sections

Guidance for completion on reverse

Date ___/___/___

Section 1a: Red Flags – If present, be alert for unmet palliative care need:	**Y**	**N**	Directly Managed	Refer to other team member	Refer SPCS
Red flag symptoms					
Clinical evidence of right heart failure					
Deteriorating Performance Status					
Has the patient or carer had repeated unscheduled contact with hospitals?					
Failure to attend clinic today?					
Section 1b: Priority referral for further assessment:	Y	N	Directly Managed	Refer to other team member	Refer SPCS
No carer?					
Patient or carer request referral to SPCS?					
You require assistance of SPCS?					

Comments:_____

Section 2: PATIENT WELLBEING ***("Does the patient have……")***	**Level of Concern**			**Action Taken**		
Who provided this information? Patient ☐ Carer ☐ Both ☐	None	Some/ Potential	Significant	Directly Managed	Refer to other team member	Refer SPCS
Unresolved physical symptoms *(including SOB/cough/ mucous; leg oedema, heartburn/reflux/poor appetite, fatigue/insomnia/daytime drowsiness, constipation, pain, cognition, voice, sore mouth, mobility, self-image or sex)?*						
Unresolved psychological symptoms / loss quality of life?						
Problems with daily living activities?						
Spiritual or existential concerns *(issues about the meaning of life and suffering)*						
Work, financial or legal concerns?						
Health beliefs, cultural or social factors making care delivery complex?						
Information needs: Prognosis ☐ Diagnosis ☐ Treatment options ☐ Financial/legal issues ☐ Support services (social/emotional issues) ☐						

Comments:_____

Section 3: ABILITY OF CARER OR FAMILY TO CARE FOR PATIENT ***("Is the Carer / Family……")***	**Level of Concern**			**Action Taken**		
Who provided this information? Patient ☐ Carer ☐ Both ☐	None	Some/ Potential	Significant	Directly Managed	Refer to other team member	Refer SPCS
Distressed about the patient's symptoms?						
Having difficulty providing physical care?						
Having difficulty coping with the patient's psychological symptoms?						
Concerned about financial or legal issues?						
Experiencing problems that are interfering with inter-personal relationships or functioning, or is there a history of such problems?						
Information needs: Prognosis ☐ The diagnosis ☐ Treatment options ☐ Financial/legal issues ☐ Support services (social/emotional issues) ☐						

Comments:_____

Section 4: CARER/FAMILY WELLBEING "Carer or family experiencing……."	**Level of Concern**			**Action Taken**		
	None	Some/ Potential	Significant	Directly Managed	Refer to other team member	Refer SPCS
Unresolved psychosocial problems or feelings *(loneliness, depression, anxiety, frustration)* that are interfering with their wellbeing or functioning?						
Grief over the future death of the patient?						

Comments:_____

IF FURTHER ASSESSMENT REQUIRED, PLEASE COMPLETE THIS SECTION:

Referral to (Name)…… ……..…….. **Patient aware of referrals**: Y N

Speciality: ILD Nurse Specialist ☐ Spiritual Care ☐ Psychology ☐ OT PT ☐ **Copy to GP** Y N

Social Services ☐ Specialist Palliative Care ☐ Local/specialist ILD Clinic ☐ Other ☐

Priority of assessment needed: Urgent (within 24 hours) Semi-urgent (2-7 days) Non-urgent (next available) ☐

Referral from: Name…… Position…….. Signature ……..

management, disease-stabilizing care, patient-centered management, and end-of-life care [35••]. PC includes all of the interventions aimed to improve and optimize qualify of life (QoL) in patients affected by progressive disease [36]. PC also includes helping the patient and their caregivers with advance care planning through the process for preference of end-of-life care [11]. Addressing goals of care early in the disease trajectory is associated with improved patient outcomes and reduced intensity of end-of-life care [37].

Palliative care can be delivered by a member of the clinical care team, referred to as primary palliative care, or an interdisciplinary team, referred to as secondary or specialty palliative care. Optimal PC for patients with chronic lung disease, such as ILD, should incorporate both primary and specialty PC [38]. Challenges facing specialty palliative care include increased demands on limited resources [39]. Supportive care is a term that has been associated with better understanding and more favorable impressions than palliative care [40]. Hospice is different from palliative care and should be offered when the patient is not expected to live greater than 6 months [41]. Resources are available to find non-hospital-based [42] and hospital-based palliative care programs [43].

The goals of PC are to prevent and relieve suffering, support the best quality of life for patients facing serious illness and their caregivers, and encourage discussions regarding EOL preferences. Studies have reported that even when patients and their caregivers understand the terminal nature of their disease, they did not appreciate that symptoms could escalate rapidly, resulting in death [30]. Because of the unpredictable nature of ILD, especially IPF, early introduction of PC should be considered a standard of care. The mantra "It is wise to hope for the best, but it is also wise to prepare for the worst" can introduce the concept of advance care planning to the patient and their caregiver [44].

Tools to Help Clinicians Identify Palliative Care Needs

Identification and management of patients' palliative care needs can be challenging for clinicians. The needs assessment tool: progressive disease in interstitial lung disease (NAT:PD-ILD) is a single page guide to prompt clinicians to assess patients' well-being, informal carers' need, and information needs prompting referral for specialty palliative care [45]. See Table 1.

Role of ILD Clinical Nurse Specialist

Nurses are considered to be central to health care provision and highly valued by patients [46]. Symptom management and palliative care are the hallmarks of nursing, especially within the realm of the ILD Clinical Nurse Specialist [47]. The CNS provides expert knowledge and advice to patients and their families throughout all stages of care and is frequently the main clinical contact for healthcare working in concert with clinical care team. The main focus is on managing symptoms and frequently adjusting the plan of care as the disease progresses. Nurse and interdisciplinary-led research makes important contributions to the evidence base of clinical practice, especially in the non-pharmacological approaches to symptom management. These collaborations work to advance the care provided for patients.

Conclusion

ILDs are complex. Education and clarity of communication are essential for the patient and their caregiver(s) to understand and appreciate the magnitude of the disease. Participating in support groups and research studies offers the patient the ability to actively manage their disease. Different ILDs can have similar symptoms and complications, and it is important to address them with symptom-specific treatments including pulmonary rehabilitation and supplemental oxygen therapy. Because the disease course is unpredictable, it is important to initiate palliative care early after diagnosis to optimize symptom management and address advance care planning. These nonpharmacologic therapies are crucial in supporting the patient and their caregiver(s) as they live with their interstitial lung disease.

Compliance with Ethical Standards

Conflict of Interest Kathleen Lindell declares no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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- •• Of major importance

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