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REVIEW

Palliative Care and End-of-Life Decisions in Interstitial Lung Disease

Rajani Surendar Bhat[1] · **Sunitha Daniel**[1]

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Abstract

Purpose of Review This review examines recent pulmonary and palliative medicine literature to evaluate the current state of palliative and end-of-life care in interstitial lung diseases (ILDs), addressing unmet needs, challenges, and evidence for best practices.

Recent Findings Patients with ILD and their caregivers do not receive timely and comprehensive supportive and palliative care due to barriers in initiating palliative care on the part of healthcare teams as well as patients and caregivers. Multidisciplinary teams with early integrated palliative care can offer better palliative care for patients with ILD and caregivers, facilitating better advance care planning, improved symptom control, and health-related quality of life.

Summary ILDs are associated with a high symptom burden for patients and a negative health impact on caregivers as well. Adoption of simple prognostication tools in clinical practice, additional training in essentials of palliative care, effective and safe non-pharmacological and pharmacological means of alleviating common symptoms, multidisciplinary care models, and telehealth are promising ways to address the gap between guidelines and implementation.

Keywords Interstitial lung disease · Palliative care · Breathlessness · Advance care planning · End of life care

Introduction

Interstitial lung disease (ILD) is an umbrella term referring to a number of lung diseases characterised by inflammation and fibrosis of the lung parenchyma to varying degrees, leading to decline of lung function and consequent morbidity and mortality. The classification of ILD has evolved with the growing knowledge of characteristic radiological and pathological features which, when combined with clinical information, allow clinicians to categorise the ILDs broadly as idiopathic pulmonary fibrosis (IPF) or non-IPF ILDs. This distinction is important as among all the ILDs, IPF is associated with rapid decline and imminent mortality within 3–5 years of diagnosis. The use of anti-fibrotics like pirfenidone and nintedanib can slow down progression, but the definitive treatment remains lung transplant which few patients can avail of [1]. Some non-IPF ILDs, which are also progressive fibrosing ILDs (f-ILDs), carry a grim prognosis just as IPF does [2]. Immunomodulatory drugs and corticosteroids can be used in some ILDs and the efficacy of combination regimens with antifibrotics is under investigation [1]. Patients with progressive f-ILD have to cope with a high disease-associated symptom burden—mainly breathlessness, cough, fatigue, and poor health-related quality of life (HRQOL) as well as depression and anxiety. The pharmacological therapies available for ILD also have associated side effects which contribute to the patient's overall symptom burden [3•]. Patients encounter exacerbations of ILD requiring increased use of emergency care, hospitalisations and supplementary oxygen support, assistance with daily activities, and require actively engaged caregivers as disease progresses. ILD patients and their caregivers, therefore, have high palliative care needs [4]. The high cost of specialised care, investigations, and treatment in low- and middle-income countries (LMIC) places an additional burden. However, low-cost interventions are available to provide palliative care for this vulnerable group [5].

✉ Rajani Surendar Bhat
rajani.bhat@gmail.com

Sunitha Daniel
hysd11@hyms.ac.uk

[1] Department of Pulmonology, Sparsh Hospitals, Bangalore, Mysore Road, Bangalore, Karnataka 560098, India

Palliative Care in Interstitial Lung Disease—Current Guidelines and Practice

Though clear international guidelines are not available, official position statements for management of IPF have included palliative care to be adopted alongside curative measures for over a decade [6]. Recently, a multidisciplinary international working group outlined best palliative care practices in ILD with the aim of 'living well' with ILD [3•]. However, a practice bias persists towards incorporating palliative care measures only at the terminal stage, or at the end of life. The Danish respiratory society (DRS) position paper on palliative care for patients with non-malignant advanced respiratory diseases addresses this misconception and lists several features that should alert the clinician to an ILD patient's need for palliative care including recurrent hospitalisations, peripheral oxygen saturation < 88%, forced vital capacity (FVC) < 50% predicted or a drop in predicted FVC more than 10%,or drop in predicted diffusion capacity (DLCO) more than 15% or a reduced 6-min walk distance (6MWD) less than 212 m. The presence of pulmonary hypertension and progressive or new onset of comorbidities should be taken into account for greater need of palliative care [7].The consensus statement on the management of ILD issued by the Indian chest society and national college of chest physicians addresses the need for incorporating palliative care in advanced ILD to alleviate distressing symptoms of dyspnoea, cough, and depression. The appropriate use of supplemental oxygen, opioids, and multidisciplinary palliative care including pulmonary rehabilitation to improve quality of life is advocated [8].

Despite growing evidence of benefit, palliative care for ILD remains under-utilised [3•]. Patients with ILD are referred for specialist care late in the course of disease when their dyspnoea is at medical research council (MRC) grades 4–5, when they are frail and unable to leave their homes due to high symptom burden [9]. They do not receive the same degree of palliative care as patients with lung cancer [10]. Opioids are either not prescribed or prescribed too late [11], and unlikely to be prescribed by non-palliative care physicians. A retrospective review of f-ILD patients in 2 Australian institutions with access to specialist palliative care (SPC) showed that despite their physicians recognising the clinical decline and 57% of patients having MRC grades 3–4 dyspnoea with chronic breathlessness, only 25% of patients had been referred for outpatient palliative care or received symptom palliation for chronic breathlessness. Forty-nine percent of patients were referred to SPC only on their terminal admission, on an average 1 day prior to death and the patients received opioids on a median of 2 days prior to death and benzodiazepines 1 day prior to death [11]. Practice surveys in various countries reveal a similar trend and indicate an awareness of the need to measure, train, and improve implementation of the rapidly growing consensus on providing palliative care to patients with ILD.

Barriers to Palliative Care Referral and Implementation of Integrated Palliative Care for Patients with ILD

Barriers to palliative care referral exist on the part of the healthcare systems as well as patients and caregivers. The lack of clear guidelines for referral criteria is one reason that there is variability in practice even in settings that have access to specialised palliative care.

A review of studies evaluating referral criteria for palliative care for patients with ILD identified hospital utilisation, functional decline, and disease progression as triggers for referral [12]. Increased hospital utilisation could be for exacerbation or other reasons, but it serves as a surrogate for greater healthcare needs. In ILD, the trigger for referral to palliative care by the primary clinician remains more disease-based than needs-based, as patients usually have had symptoms for considerable time before referral to palliative care [13].

A Danish national survey following the DRS position paper provides insights into the challenges faced even by clinicians with a positive attitude and knowledge of palliative care in ILD in implementing these practices. Seventy-one percent of clinicians surveyed had knowledge of the DRS position paper. Lack of time, as reported by 63%, and inadequate staff for multidisciplinary care, reported by 52%, point to systemic changes needed to provide timely palliative care for patients with ILD [13]. The uncertain trajectory of disease and difficulty in prognostication were also identified by 63% as barriers to addressing palliative care and advance care planning (ACP) conversations. An evaluation of barriers and facilitators to best care for IPF in Australia identified shortage in psychological support services and regional inequity in specialised IPF services [14, 15]. In addition to these factors, a survey of 68 Pulmonary Fibrosis Foundation (PFF) care centre network sites suggests that while ILD specialists maybe able to provide primary palliative care services for their patients, there is a need for increased access to SPC referral, along with standardisation of assessment of symptom burden, HRQOL, and timing of SPC referral to improve palliative care in ILD [16]. Another recent survey across respiratory and palliative care professionals shows that the common reasons for referral are optimisation of symptom control, providing psychosocial and spiritual support needed with the patient's decline, and end of life care [17]. Furthermore,

specialists in life-limiting chronic respiratory diseases like cystic fibrosis may be overestimating their competence at providing advanced palliative care, especially end of life care [18].

On the patients and caregivers' side, cultural factors play a role as clinicians delay ACP discussions due to real or perceived unwillingness for difficult conversations [15]. Patients and caregivers may perceive referral to SPC as loss of hope and abandonment. Therefore, the timing of referral is a challenging issue to address; too early may discourage patients and caregivers and too late does not allow the palliative care team adequate time to build a relationship of trust and facilitate optimal symptom control and advance care planning.

Unmet Needs—Patients and Caregivers

The existing challenges in providing palliative care for patients with ILD lead to a gamut of unmet needs in patients with ILD and their caregivers. Involving all stakeholders in world-café style discussions in qualitative research, to understand the care needs of people living with f-ILD, has shown avenues for improvement in patient-centred care integrated palliative care [19]. These unmet needs range from disease information, involvement of caregivers in care-plan discussions, symptom control, appropriate oxygen and opioid prescription, pulmonary rehabilitation, uncertainty on how to manage exacerbations and emergencies, advance care planning, and lack of opportunity to access lung transplant or enrolment in clinical trials. Lack of acknowledgement of spiritual needs, psychological support, and legal guidance for advance directives emerged as areas for patient advocacy groups to focus on systemic solutions. These unmet needs extend to the support needed during bereavement of caregivers who lose loved ones to ILD [20]. As we design and study the impact of palliative care interventions to address the unmet needs of patients and caregivers, it must be kept in mind that the current literature on palliative care in ILD is lacking in standardised instruments to measure both needs and outcomes. A combination of tools like the King's brief ILD questionnaire (kBILD) or the palliative care needs assessment tool in ILD (NAT PD ILD), the Leicester cough questionnaire (LCQ) maybe required to get a complete picture of a patient's needs [20]. Additionally, the role of palliative care screening tools similar to the Edmonton symptom assessment scale (ESAS) which rates the intensity of nine commonly experienced symptoms on a numeric scale, that have been validated in patients with cancer, could be studied to screen for a wider range of symptoms regularly [21]. Patient-related outcome measures (PROM) in the physical, mental, social, and spiritual health, and HRQOL dimensions to address these unmet needs are key to better design of patient centred specialised ILD clinics 2 [22, 23]. A further step would then be to assess patient reported experience measures to assess quality of palliative care in ILD [20].

Burden of Symptoms in ILD

A systematic review on the symptom prevalence of patients with ILD has shown that breathlessness, cough followed by heartburn, and depression are the most common symptoms. The other symptoms include fatigue, reduced exercise capacity, anxiety, poor sleep, and pain [24]. Table 1 below shows that the high burden of symptoms in patients with ILD is comparable to the experience of patients with life-limiting illnesses like cancer, AIDS, etc.

A key tenet of palliative medicine is to assess and address all the aspects of a patient's suffering. This concept of 'total pain-physical, psychological, social and spiritual' can be applied to assessment of dyspnoea as 'total dyspnoea' in patients with ILD [24], where we address all factors contributing to the breathlessness experienced by the patient. In addition to optimisation of disease-directed therapy and therapy for comorbidities, chronic refractory breathlessness is best managed with a multi-modality approach. The breathing-thinking-functioning model [27] aids in the optimal approach to management of 'chronic breathlessness syndrome' [28]. For most patients with f-ILD, their dyspnoea despite optimal medical treatment of the disease leads to severe debility and breathless interventions with a syndromic approach can serve their palliative care needs better. Cough is a particularly distressing symptom to both patients and caregivers. Caregivers experience emotional burden with the patient's dyspnoea and cough. The negative feelings evoked in caregivers due

Table 1 Summary of the prevalence of symptoms in Cancer, AIDS, CHF, COPD, ESRD and progressive f-ILD (figures for other conditions taken from Solano et al. 2006) [25, 26]

Symptoms	Progressive f ILD	Cancer	AIDS	CHF	COPD	ESRD
Pain	9%	30–94%	30–98%	14–78%	21–77%	11–93%
Depression	10–49.2%	4–80%	17–82%	6–59%	17–77%	2–61%
Anxiety	22–58%	3–74%	13–76%	2–49%	23–53%	7–52%
Fatigue	7.6–29%	23–100%	43–95%	42–82%	32–96%	13–100%
Breathlessness	54.7–98%	16–77%	43–62%	18–88%	56–98%	11–82%
Insomnia	6–46.6%	3–67%	40–74%	36–48%	15–77%	1–83%
Nausea	13%	2–78%	41–57%	2–48%	4%	8–52%
Diarrhoea	2%	1–95%	29–53%	12%		8–36%

to the patient's relentless cough also can lead to feelings of guilt in the caregiver [29]. Fatigue is another symptom that is hard to measure, with physical and mental components. A proposed model to manage fatigue is to assess (i) predisposing factors such as biological vulnerability and lack of support, (ii) precipitating factors such as the underlying ILD, acute worsening, and social or psychological stressors, and (iii) perpetuating factors such as physical inactivity, poor coping mechanisms, and poor quality or disrupted sleep architecture [30]. Patients with ILD experience poor sleep, some of them may have sleep disordered breathing like obstructive sleep apnoea as comorbidities worsening their symptoms. Insomnia is another complaint with cough and gastro-oesophageal reflux being a contributing factor for some patients [31]. Dyspnoea and cough have a debilitating impact on the overall quality of life of patients with ILD and also are associated with depression and anxiety over time. There is an association between dyspnoea and the need for supplemental oxygen and anxiety [32].

A good understanding of symptom clusters will help to screen patients for other common symptoms when a diagnosis of ILD is made and provide better overall symptom management.

Management of Common ILD Symptoms

In this section, we elaborate on the evidence for management of the common symptoms of breathlessness and cough. Higginson et al. demonstrated that well-designed breathlessness intervention services have benefits in the form of mastery of breathlessness and improved survival with early integrated palliative care in patients with chronic respiratory disease when compared with controls [33]. Though this study is limited by the small number of patients with ILD, it provides a model for incorporating early integrated multidisciplinary palliative care with a goal of 'mastery' of symptoms rather than resolution of symptoms, and 'living well' with ILD.

Non-Pharmacological Measures for Breathlessness

Oxygen

The recent ATS clinical practice guidelines for home oxygen therapy for patients with chronic lung disease make a case for the use of oxygen in some patients with ILD with a strong recommendation for the use of oxygen in patients with resting hypoxia and a conditional recommendation for use in patients with exertional desaturation (albeit based on low quality evidence) [34]. While there may be no significant benefit in improvement of dyspnoea, improved exercise capacity and 6MWD can have a positive impact on quality of life.

Pulmonary Rehabilitation

Recent large multicentre studies on the impact of pulmonary rehabilitation in patients with ILD confirm the findings of prior studies of improvement in 6MWD and exercise capacity. A reduced perception of fatigue was also noted in the exercise group. While there was a distinct loss of benefit after cessation of the pulmonary rehabilitation program, a promising 28% of patients had sustained gain in 6MWD on follow up [35].

Miscellaneous

Cool air to the face through a handheld fan is useful for episodic worsening of breathlessness [3•]. Cognitive behavioural therapy has been recommended for symptoms related to sleep and disruptive coping mechanism. Multimodality speech pathology training, cough suppression techniques, and vocal hygiene are helpful in chronic cough [36]. Acupuncture and music therapy do not have sufficient evidence beyond case reports to support their use for breathlessness in ILD [37].

Pharmacological Measures for Breathlessness and Cough

Opioids

The role of opioids in chronic breathlessness due to chronic obstructive pulmonary disease is well supported. However, there is not enough high-quality evidence to support the use of regular low dose opioids for chronic breathlessness in ILD. Recent studies of the use of low dose morphine for chronic breathlessness suggest a trend towards benefit but no significant improvement in dyspnoea scores in the morphine group [38]. However, the same study also notes an improvement from baseline dyspnoea and 6MWD in the hour following administration of morphine, suggesting a possible role for low dose opioid in episodic breathlessness and improved QOL with improved 6 MWD. Small case series suggest benefit with buccal hydromorphone for breathlessness in patients with ILD with a structured breathlessness support algorithm; however, more evidence is required for the buccal route [39]. Opioids find a place in the chest guidelines and expert panel report for the treatment of ILD-associated intractable cough without high-quality evidence as an option to alleviate high symptom burden in patients with ILD, just as in unexplained cough [36]. The adage of 'start low and go slow' applies to the use of opioids

in patients with chronic respiratory disease [40]. Safety of use of low dose opioids has been established with no reported increase in mortality with the use of opioids [41]. Constipation and nausea are reported with opioids and should be managed concurrently.

Benzodiazepines

Low dose benzodiazepines can be used to treat anxiety associated with chronic breathlessness and in conjunction with low dose opioids for relief of dyspnoea in end-of-life care (EOLC). No increase in mortality has been noted with low dose benzodiazepines or low dose opioids in patients with ILD, with no increase in hospitalisations as well [41]. There is a dose association of benzodiazepines with higher mortality when used in higher doses [42].

Antifibrotics and Immunomodulators

The commonly discussed outcomes of studies on antifibrotics have been the measurable improvement in FVC or 6MWD. However, post hoc analysis of the large studies on antifibrotics shows that both pirfenidone and nintedanib have demonstrated some benefit in reducing worsening of breathlessness [43, 44] in patients with IPF and f-ILD, as well as improvement in objective measures of cough after use beyond 1–2 years [45, 46]. In patients with CTD-ILD, immunomodulatory drugs such as mycophenolate mofetil and cyclophosphamide have shown improvement in patient reported outcomes of breathlessness and cough [47].

Other Medications

Diuretics, bronchodilators, and corticosteroids may be used to palliate symptoms based on clinical assessments in conjunction with other medications for symptom relief. A feasibility study of mirtazapine for severe breathlessness showed tolerability and safety and further multicentre studies are awaited [48].

The current recommendations for management of cough in ILD do not support the routine use of proton pump inhibitors in the absence of gastroesophageal reflux disease, nor are inhaled steroids recommended for cough in sarcoidosis. Neuromodulators and inhaled cromolyn sodium hold promise as potential therapies. Cough remains a difficult to treat symptom with inadequate supportive evidence for pharmacological therapies.

Symptoms Due to Medications for ILD

A recent case control study raised concerns about an association between the use of selective serotonin and norepinephrine reuptake inhibitors (SSRI/SNRI) used for treating depression in the elderly and occurrence of ILD. As patients with ILD may have concomitant depression, patients already on these medications should be monitored for worsening respiratory symptoms [49].

Both antifibrotics and immunomodulatory drugs are known to cause a number of undesirable side effects [1, 44]. Pirfenidone is known to cause anorexia, fatigue, and nausea and nintedanib can cause diarrhoea. The symptoms can be distressing to a level where patients are unable to take the recommended therapeutic dose or may develop new conditions like infectious complications with immunosuppression. A clinician must therefore screen for symptoms regularly at each visit and assess for trends. A combination of screening tools like kBILD and those used in monitoring symptom burden, like the ESAS used in the palliative care of patients with cancer, could provide clinicians with a way of assessing when the treatment is potentially causing greater distress to the patient than the underlying disease. In the absence of evidence or consensus guidelines to aid the clinician's dilemma in these situations, the ethical principle of autonomy, i.e. following the patient's expressed wishes and goals of care, is the deciding factor for whether a medication should be continued.

Advance Care Planning and End of Life Care Decisions

Prior reviews of studies on advance care planning (ACP) in chronic respiratory disease have brought to light the lack of timely ACP discussions with patients and caregivers. The uncertainty of prognosis in non-malignant advanced lung disease is often quoted as the challenge in ascertaining the right timing of initiation of ACP [50]. Perceptions of the clinical team of lack of readiness on patients' and caregivers' part and the perception of caregivers of clinical team's hesitation in initiating difficult conversations points to a need for increased training in communication. Recurrent themes that emerge from qualitative studies and focus group discussions on ACP in patients with ILD are (i) conversations occur late, (ii) insufficient information—more information conveyed simply is better, (iii) clarity on important fears, and (iv) reassurance of support at the end of life with options provided. Comforting safety, good team support, balancing honesty with information,

and repeated conversations at 'turning points' in disease trajectory were identified as patient's needs in ACP.

While clinicians often state the uncertainty of disease prognosis as a limiting factor in initiating ACP, a recent study on the validity of a staple tool in ACP—the surprise question (SQ) shows its utility in IPF as well. The SQ requires the clinician to ask themselves a simple question with no additional data.The question is 'Would you be surprised if this patient were to die within the next 1 year?' Pulmonologists and specialist ILD nurses identified patients likely to die within 1 year with a sensitivity of 68% and specificity of 82% in a prospective study of 140 patients with IPF [51•], outperforming another prognostic indicator, the GAP score, in predicting 1-year mortality.

Including the SQ in checklists in routine ILD clinic visits can therefore be an important trigger to initiate ACP. However, ACP must not be relegated to an item on a checklist as it is a process that can take place over multiple visits with different goals for each discussion. This should include discussions about initiation of invasive life-sustaining treatment like mechanical ventilation, haemodialysis or extracorporeal membrane oxygenation, artificial nutrition, and do-not-attempt-resuscitation (DNAR) orders. In patients admitted to the ICU with acute exacerbation of ILD, the need for mechanical ventilation has been shown to be associated with higher mortality irrespective of the type of ILD [52]. Appropriately timed early ACP with documentation and communication with all care providers allows for goal-concordant care for patients who are not being considered for lung transplant, for whom invasive life-sustaining therapies may not be beneficial.

Lack of involvement of palliative care in patients with ILD can contribute to patient spending their last days of life in acute care settings, with high investigation and treatment burden [53, 54]. Due to the unpredictable nature of the disease, patients may experience acute exacerbations before ACP has been initiated. Previous exacerbations, higher oxygen requirement, and poor quality of life prior to hospitalisation can aid in decision-making for those who will not benefit from intensive care measures.

Interestingly, early involvement of palliative care for patients with ILD with dyspnoea has shown greater uptake of ACP, more use of opioids for dyspnoea but also shown higher median survival in the palliative care intervention group, with lower ICU admission despite higher initial symptom burden [55, 56]. This finding supports integrated palliative care for patients with ILD wherever palliative care services are available.

Foregoing Life Sustaining Treatment and Palliative Sedation

The WELPICUS study established consensus among 32 countries participating on matters related to informed consent, withdrawing and withholding life sustaining treatment, shared decision-making, and palliative care for ethical EOLC in ICU [57]. Withdrawal and withholding life-sustaining treatment are considered the same on ethical principles, though there is variability in practice across different countries. Palliative care for symptom relief should be standard of care for all hospitalised patients with ILD in all settings including the ICU where de-escalation of medical support is contemplated. For those patients in a respiratory crisis with ILD who receive their EOLC in hospital, oxygen via high flow nasal cannula (HFNC) therapy is useful as patients do not mandatorily require the ICU setting for HFNC. A bereavement survey of caregivers of deceased patients shows that over 98% of patients who received HFNC were not in ICU and the caregivers reported higher scores on the good death inventory (GDI) and quality of death and dying (QODD) with this respiratory modality [58, 59]. Palliative sedation as part of EOLC for patients with ILD follows the doctrine of double effect. It is to be used for patients with terminal refractory breathlessness, and during withdrawal from invasive life-sustaining treatment or HFNC. The primary goal in the use of opioids and/or benzodiazepines used transdermally or as continuous subcutaneous or IV infusions is optimal titration to achieve relief of symptoms. The secondary effect of the medication may lead to hemodynamic or respiratory decline; however, the primary intention is relief of distressing symptoms. There is no deliberate attempt to hasten the dying process [60].

The decision of the right place for the patient at the end of life should be guided by the patient's expressed wishes. Home and hospice care for end of life in ILD is feasible [61].

Lung Transplant

The complex palliative care needs of patients with ILD who are lung transplant candidates, recipients, and their caregivers would be best served by integrated or embedded palliative care services as a part of the transplant team. Utilisation of recently validated tools such as the lung transplant quality of life tool (LT QOL) may provide more information about the novel needs of transplant patients and their caregivers. While there is limited evidence in the domain of lung transplant, evidence from studies involving bone marrow transplant and other end-organ failure solid

organ transplant patients suggests that integrated palliative care in the peri-transplant team with variable frequency of engagement based on needs assessment may improve patients' quality of life [62].

Caregiver

Being the caregiver of a patient with ILD can impair HRQOL of the caregiver, placing burdens on emotional health. Palliative care for ILD patients includes education and support offered to caregivers for coping and resilience. Respite admissions to hospital or hospice can be used on occasion for relieving caregiver stress.

Qualitative studies of spousal bereavement after death due to ILD suggest that the timing of death, place of death, and the process of death influence grief and bereavement. Partners and caregivers of patients with f-ILD are at risk for prolonged grief rather than a normal grief and bereavement response [59], further impairing their HRQOL after the death of the partner.

Integrated Palliative Care Models

The positive impact of integrated palliative care for patients with ILD extends from early days of diagnosis and management to disease progression, end-of-life-care, and bereavement [33, 61].

Palliative care co-management of patients with ILD was shown to increase ACP notes to 100% with completed advance directives increasing from 22.6 to 35.5%, along with completed Physician orders for limitation of life-sustaining treatment increasing from 0 to 35.5%. Greater use of short acting opiates for relief of episodic dyspnoea was observed. Telemedicine was used to facilitate palliative care co-management in 48.4% of patients [63•]. Involvement of psychologists and palliative care in a multidisciplinary team meeting with ILD was also associated with greater ACP and resolution of patients' unmet needs [64]. Nurse-led facilitated ACP discussions are effective and acceptable to patients with ILD and can help improve patients and caregivers' knowledge and preparedness [65, 66]. The Munich Breathlessness Service offered a multidisciplinary breathless support program similar to CBIS where patients were recruited for a study through media announcements [67].

In an increasingly digitally connected world, social media and telehealth offer opportunities to expand care models and create hybrid models to improve access to early integrated palliative care.

Future Avenues for Research

The review by the Lancet international working group on palliative care in ILD, in addition to outlining a comprehensive approach to the ABCDE of ILD (assessment, backing, comfort, disease modification, and end of life care), also highlighted the barriers and challenges to implementation [3•]. These offer insights into future areas of research for the impact and cost effectiveness of palliative care in ILD. Due to the interconnectedness of the physical, mental, social, and spiritual dimensions of suffering in patients with ILD, there is a need to incorporate patient-related outcome measures and patient-related experience measures in future studies [68].

Key Recommendations

1. Increase training of ILD clinical teams in palliative care principles and communication.
2. Offer patients an extra layer of support with early integrated palliative care in ILD for appropriate symptom management, ACP, and end of life care plans.
3. Consistently assess symptoms, HRQOL, and PROM using existing validated tools on each visit to understand an individual patient's disease trajectory.
4. Advocate competency in non-pharmacological symptom relief measures and safe use of opioids for breathlessness in various settings to offer multimodality breathlessness support services.
5. Multidisciplinary team approach with enhanced use of telehealth to improve access to pulmonary rehabilitation, palliative care, psychologists, respiratory therapists, and social workers where available.
6. Caregiver support.

Compliance with Ethical Standards

Conflict of Interest The authors declare no competing interests.

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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