



Palliative care for patients with pulmonary fibrosis: symptom relief is essential

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Patients with PF-ILD suffer from heavy symptom burden and poor quality of life. Early use of opioids and low dose BDZ in severely ill patients with respiratory compromise may lead to improved symptom control and quality of life for this patient group. <http://ow.ly/x50e30mJ6ko>

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Symptom burden plagues patients with progressive fibrotic interstitial lung disease (PF-ILD) [1]; this symptom burden, in combination with poor prognosis and uncertainty of disease course heavily impacts quality of life, both for patients and family members [2, 3]. Patients with PF-ILD are confronted with a wide range of diagnoses and prognoses; some may live many years with a disease that is responsive to treatment, while others with PF-ILD often have a disease trajectory similar to and/or worse than that of lung cancer [4, 5].

Regrettably, treatment interventions targeting the cascades of the pathways of lung tissue response as a consequence of injury, inflammation and fibrosis to date have not lessened this burden. As a consequence, patients with PF-ILD are confronted with progressive symptoms and the overall burden worsens while they endure the suffering in their shortened life span. Indeed, significant emotional sufferings that include depression, anxiety and at times panic episodes are unfortunate expected consequences that confront most, if not all patients with irreversible and progressive fibrotic interstitial lung disease.

Patients suffer common symptoms related to their chronic lung disease which contributes greatly to their morbidity and impacts their quality of life [1]. Symptoms patients most frequently experience include dyspnoea, cough, fatigue, anxiety and depression, and patients experience varying degrees of symptom burden and functional decline as their PF-ILD progresses [1, 2]. In a review of patients with PF-ILD, the overwhelming majority of patients reported breathlessness (68.2–98%), cough (59–94%), heartburn (25–65%) and depression (10–49%); this review also found that patients reported constitutional symptoms including sleep disturbances, fatigue, weight loss, and anorexia [1].

It is critical that the clinician is able to make a confident diagnosis of the specific form of PF-ILD as this will be useful to formulate patient-centred, personalised counselling and a plan of care to achieve remission or stabilisation of the disease process, when possible, with identification of symptom burden and repeated assessment throughout the disease course [6]. Delivery of information to patients and caregivers regarding the magnitude of the PF-ILD diagnosis needs to be done in a caring, empathic manner to

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convey realistic hope to the patient and their caregiver, as the delivery of an PF-ILD diagnosis is devastating and forever changes their lives [7]. Early educational programmes can help to increase knowledge of the disease and management options so that patients and their caregivers can have a better understanding of the effect and consequences of these relentlessly progressive diseases [8, 9].

Once symptom burden is accurately assessed, there are a number of pharmacological and nonpharmacological therapies available to reduce symptom burden [6, 10, 11]. These include regular counselling, anxiolytic and anti-depressive medications, low-dose narcotics, pulmonary rehabilitation, supplemental oxygen therapy, and early referral to palliative care to alleviate symptom burden and address advance care planning, with the ultimate goal of improving quality of life [3, 12, 13].

In this issue of the *European Respiratory Journal*, BAJWAH *et al.* [14] report the results of a population based longitudinal cohort study of PF-ILD patients starting long term oxygen therapy in Sweden between 2005 and 2014. They examined association of benzodiazepines and opioids on risk of admission to hospital and death in this cohort of 1603 patients and found that benzodiazepines were used by 196 (12%), and opioids by 254 (15%) patients when they were initiated on long term oxygen therapy. They reported no association between benzodiazepines and increased admission, but treatment with higher dose *versus* lower dose benzodiazepines showed association with increased mortality. It is important to note that these patients were advanced in their disease course and some of these patients were receiving benzodiazepines for end of life management of anxiety-related breathlessness. Benzodiazepines reduce the sensation of breathlessness through decreasing the anxiety associated with breathlessness and should be considered at lower doses to decrease symptom burden. Their findings also showed that depressed/anxious patients were much more likely to be prescribed benzodiazepines and opioids. Low or high dose opioids showed no association with increased admission or increased mortality in this cohort. This is the first ever study to look at safety concerns of these pharmacological treatments for symptom management in patients with PF-ILD, and their findings suggest that when benzodiazepines and opioids were used, they were not used until the disease was advanced and the patients were oxygen dependent. These findings support the early use of opioids and low dose benzodiazepines in severely ill patients with respiratory compromise. Improvements in breathlessness management early in the disease course may lead to improved symptom control and quality of life for this patient group.

In essence, patients with PF-ILD, including advanced IPF, suffer from heavy symptom burden and a poor quality of life [15]. This calls for comprehensive symptom management and integrated palliative care concomitant with disease-modifying therapies as soon as the patient is diagnosed with the PF-ILD. The incorrect perception that palliative care is synonymous with end of life care, with no role earlier in the course of ILD, has created a culture of neglect [12]. The stigma of prescribing opioids and benzodiazepines contributes to the symptom burden that patients unnecessarily suffer [12, 16].

Early integrated palliative care for patients with lung cancer has shown substantial benefits, such as lower depression scores, higher health-related quality of life, better communication of end of life care preferences, less aggressive care at the end of life, and longer overall survival [17].

We believe that incorporating early palliative care and regular counselling will allow patients with PF-ILD and their caregivers to obtain better and more effective pharmacological and psychosocial interventions to improve quality of life throughout the disease course. Sensitive advanced care planning, and timely patient-centred end of life care with much-needed counselling with patients and their caregivers, as well as appropriate pharmacotherapy to alleviate emotional and physical suffering, must be an integrated part of routine care of patients with PF-ILD.

TABLE 1 Some palliative care studies in interstitial lung disease

Study	Manuscript title
BAJWAH and YORKE [18]	Palliative care and interstitial lung disease
BARRIL <i>et al.</i> [19]	Palliative care in interstitial lung disease: results of a Spanish survey
KALLURI <i>et al.</i> [20]	Beyond idiopathic pulmonary fibrosis diagnosis: multidisciplinary care with an early integrated palliative approach is associated with a decrease in acute care utilization and hospital deaths
LINDELL <i>et al.</i> [9]	Randomised clinical trial of an early palliative care intervention (SUPPORT) for patients with idiopathic pulmonary fibrosis (IPF) and their caregivers: protocol and key design considerations
RUSH <i>et al.</i> [21]	Access to palliative care for patients undergoing mechanical ventilation with idiopathic pulmonary fibrosis in the United States
SHARP <i>et al.</i> [22]	Development of tools to facilitate palliative and supportive care referral for patients with idiopathic pulmonary fibrosis

Table 1 lists some of the current palliative care studies that are underway. While we are not aware of all clinical studies that are ongoing and/or have been completed, it is hoped that there are and/or will be more clinical studies addressing palliative care in this cohort of patients and provide evidence to enhance further understanding and management of PF-ILD and improve outcomes that are meaningful to these patients.

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