Palliative Care Rounds

Early and Integrated Palliative Care to Achieve a Home Death in Idiopathic Pulmonary Fibrosis

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Introduction

Idiopathic pulmonary fibrosis (IPF) is the most common idiopathic interstitial pneumonia characterized by progressive fibrosis. Chronic refractory breathlessness is the clinical hallmark and the most debilitating symptom of IPF; it is strongly associated with poor quality of life and mortality.[1,2] Dyspnea in IPF remains grossly undertreated.[3] The current IPF treatment guidelines recommend palliative care referral and a symptom-based approach as an adjunct to disease-focused care.[4] However, it is well recognized that most IPF patients do not receive palliative care during the course of their illness or at end of life.[5,6] As a result, these patients suffer from not only a poor quality of life but also a poor quality of dying and death.[7] Although most patients express the preference for a home death, unfortunately the majority die in hospitals, including intensive care units.[5] In fact, outside of a clinical feasibility trial setting, home deaths have not been described in the literature, which quotes 100% of deaths occurring in acute care.[5,8] Most patients and families believe that their symptoms at the end of life will be managed more effectively in hospital. Concurrently, respiratory specialists are not familiar with facilitating patient care and management in the home or to providing palliation of their symptoms at home.[9] In addition, it is generally believed that patients with end-stage lung disease are bed bound.[4,10]

To our knowledge, there are limited published reports describing an approach that allows patients with IPF to die at home without being bed bound. We describe a case where patient-centered multidisciplinary care provided early integrated palliative care and supported a very active end-of-life experience for a patient with IPF, including dying at home which was his preferred location of death. We believe that early integrated palliative care is key to remaining active, independent, and at home, and thereby facilitating a good quality of death and dying in IPF.

Case Report

Our patient was a 72-year-old man with advanced IPF (usual interstitial pneumonitis [UIP] pattern on high-resolution computerized tomography [CT] scan of chest) and mild pulmonary hypertension. Medical history included diabetes, hypertension, gout, obesity (body mass index 35), and gastroesophageal reflux disease. He was diagnosed with IPF one year prior and was subsequently referred to us. At initial presentation, he had mild exertional dyspnea, Medical Research Council Class 3/5, cough productive of about a teaspoon per day of clear sputum, and weekly episodes of heartburn. He was assessed by a multidisciplinary team consisting of a pulmonologist with expertise in interstitial lung diseases (ILD), a pulmonary rehabilitation medicine specialist with expertise in palliative respiratory care, a physiotherapist, a dietitian, and a nurse.

We assessed all symptoms of most concern to him, asking him to rate his dyspnea and other symptoms in the context of his daily activities on a linear scale of 0−10, where 0 indicated absence of the symptom and 10 indicated the worst severity.[11] Physical examination revealed an alert individual in mild respiratory distress at rest, with tachypnea (22/minute), SpO_2 96% on room air, and bilateral velcro crackles in the lung bases on chest auscultation, but without clinical signs of pulmonary hypertension or right-sided heart failure. Pulmonary function tests showed a low normal total lung capacity suggesting mild restrictive pattern

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(forced vital capacity [FVC] 75%; total lung capacity [TLC] 79%) and severe diffusion impairment (diffusing capacity or transfer factor of the lung for carbon monoxide [DLCO] 39%). Chest CT showed progression of IPF compared to a study done one year prior. On a six-minute walk test, he was able to walk a distance of 467 m which resulted in a lowest arterial oxygenation saturation of 80% on room air. He started supplemental oxygen: 3 LPM at rest, 9 LPM with light activity, and up to 10 LPM during exercise, aiming for a resting SpO_2 of 98%−100% before activity and nadir desaturation of 90% with exertion. He trialed an antifibrotic agent for a few weeks but did not tolerate it. He was started on daily antireflux therapy with a proton pump inhibitor. Dyspnea management strategies included attending an ILD-specific pulmonary rehabilitation program. He obtained a four-wheeled walker that was adjusted to accommodate two oxygen tanks (E cylinders). Advance care planning was initiated at the first clinic visit: he chose medical care for reversible conditions, without resuscitation or ICU care; his preferred place for end-of-life care and death was home. He and his wife attended the IPF patient support group.

At his three-month follow-up visit, he was noted to have worsening dyspnea Medical Research Council 4/5 with further decline in lung function (FVC 47%). Repeat CT chest showed no pulmonary embolism but increased ground glass in the areas of previously established fibrosis. He was hospitalized for management of an acute exacerbation of IPF, possibly precipitated by insufficient use of supplemental oxygen while hunting with his grandson. He was placed on high-dose prednisone and empiric broad spectrum antibiotics. Repeat echocardiogram showed increased RV overload suggesting worsening pulmonary hypertension. The multidisciplinary ILD team was consulted for further inpatient management, which included the following: increasing his oxygen prescription to 12 LPM at rest and 15 LPM on exertion; initiating narcotics at 0.1 mg of hydromorphone by mouth at meal times and bedtime for baseline breathlessness, preactivity buccal hydromorphone for incident (anticipated) dyspnea (0.1 mg 5−10 minutes before light activity and 0.2 mg before more intense activity) and 0.2 mg buccal every 10 minutes prn for crisis (unanticipated) dyspnea (Table 1). Before hospital discharge, a family meeting was held to discuss how this most recent deterioration marked a near-death experience and to review options for care with the next episode, especially since the patient desired a home death. A community-based home care team was assembled to support his wishes for care at home: this team was comprised of a registered respiratory therapist, nurse practitioner, palliative care nurse, occupational therapist, and physiotherapist, in collaboration with the ILD multidisciplinary physicians.

Over the subsequent few weeks, oral and buccal opioids were adjusted to meet his increasing dyspnea (Table 1). Clinical assessments were carried out at several time points by the nurse practitioner who now served as his primary care practitioner, the home care registered respiratory therapist (RRT), and the palliative home care nurse. The ILD clinic team was in constant communication with the community home care team. Goals of care were reviewed at each encounter, where the patient emphasized his wish to not be admitted to the hospital again. His goals were to visit with friends and family, enjoy dinner and wine, and remain as independently active as possible in spite of progressing IPF. With proper education and coaching, he became quite skilled in

Table 1
Dyspnea Management During the Last Two Months of Patient's Life

Months Before Death	Prescription	Activity Level	Dyspnea
2	Hydromorphone - Resting dyspnea 0.1 mg tid - Incident dyspnea 0.1−0.2 mg buccal 10-minute preactivity - Crisis dyspnea 0.2 mg buccal repeat q10 minutes till relief Oxygen 3 LPM rest 10 LPM with activity	Ambulating outside home	At rest 5/10 With activity 10/10
0.25	Hydromorphone - Resting dyspnea 0.6 mg tid - Incident dyspnea 0.6−1 mg buccal 10-minute preactivity - Crisis dyspnea 1 mg buccal repeat q10 minutes till relief Oxygen 15 LPM rest and 20 LPM with exertion	Went on road trip, visited wineries, went raspberry picking	At rest 2/10 With activity 7/10
Day of death	Hydromorphone - Resting 2 mg po qid - Incident dyspnea 2 mg buccal 10-minute preactivity - Crisis 2 mg buccal with q10 minutes repeat dosing Lorazepam 1 mg SL as needed for anxiety and agitation Methotrimeprazine 6.25−12.5 mg q4−6 hours for crisis agitation Oxygen 24−25 LPM rest and up to 30 LPM with exertion	Ambulating with some assistance within home Planning to visit a bakery for breakfast next morning	At rest 3/10 With activity 6/10

self-management of dyspnea. Within one month of hospital discharge, he went on a three-week road trip (traveling more than 2500 km in total on high flow oxygen) to visit his daughter and even went raspberry picking on 15−18 LPM of supplemental oxygen. The effective use of self-management protocols for dyspnea, adequate oxygen supplementation, and thorough patient and caregiver education, including who to call, allowed him to accomplish his goals.

He remained active and died four days after returning home from the trip. Two nights before he died, one of his two oxygen concentrators (t'd in together to supply sufficient oxygen) broke down. This oxygen desaturation precipitated a dyspnea crisis and abrupt exacerbation of his IPF. He self-managed this crisis at home with the advice and urgent home visit from his home care team as well as the ILD clinic physician; there were no 911 calls. Further adjustments to his oxygen and narcotic doses allowed him to remain independent for activities of daily living (e.g., feeding, toileting, bathing, shaving, and dressing). Additional medications prescribed at the end of life included lorazepam 1 mg sublingual every four hours as needed for anxiety with crisis dyspnea and methotrimeprazine (25 mg/mL) 6.25−12.5 mg buccal for sleep and every four to six hours prn for crisis agitation.

He died 38 days after his last hospitalization for dyspnea without using further acute care resources and living fully until his last breath: telling his grandson where to get cinnamon buns for breakfast the evening before he died; about an hour before death, he told his family that he still had 20 minutes left on his extra oxygen tank. He was not in a moribund state. After our patient died, we administered the CANHELP Lite bereavement questionnaire to our patient's wife who responded as very or completely satisfied to all questions.

Comment

Home death is preferred but rarely achieved in advanced IPF. Most IPF patients die in acute care settings.[5] This is also true in lung cancer and other chronic lung diseases such as chronic obstructive pulmonary disease (COPD). In a retrospective cohort study of patients with lung cancer 40% of deaths occurred in the hospital, 29% at home, and 17% in hospice.[12] In a Canadian study of 1098 patients who died with COPD or lung cancer recorded as the cause of death, the proportion of home deaths was only 15.4%. These decedents did not receive home palliative care services.[13] We believe that the lack of early and integrated palliative care is one of the main reasons for poor end-of-life care in IPF which leads to deaths in acute care. In this case study, we describe how our model of care achieved a home death, allowed good symptom control thereby providing an acceptable quality of life while avoiding the terminal bedbound state as described in patients with advanced lung disease.[4,10] We identify five components of early and integrated palliative care that helped achieve a comfortable home death for our patient.

The first component is advance care planning (ACP), which is integral to early integrated palliative care and a key to achieving a home death in IPF when it is the preferred location. This planning starts with an open, honest conversation between patient, caregiver, and care team. Although many respirologists may feel reluctant to engage in such a conversation, studies clearly document that patients want this discussion. ACP is an "organized, ongoing process of communication."[14,15] The reality is that ACP in IPF is not occurring at all or is happening too late in most cases.[3] As a result, most patients pass away without good symptom control, on hospital wards or ICUs.[5,6] In our case report, ACP was initiated at the first visit, as is our routine practice in our multidisciplinary ILD Clinic. Given the unpredictable course in IPF as exemplified by the more rapid decline in our patient's disease course, early conversations about ACP are imperative. Current research efforts in palliative care for IPF tend to focus on developing needs assessment tools to identify an appropriate time point for referral to palliative care by the ILD team. This approach may create an unnecessary delay in initiation of urgently needed conversations and care. In our clinic, we do not refer to a palliative care consultation service outside of the clinic. We integrate both a pulmonary rehabilitation and a palliative care approach into usual IPF care, so that the focus is on maximizing function and quality of life from the time of the very first visit. This allows for early symptom management which is an

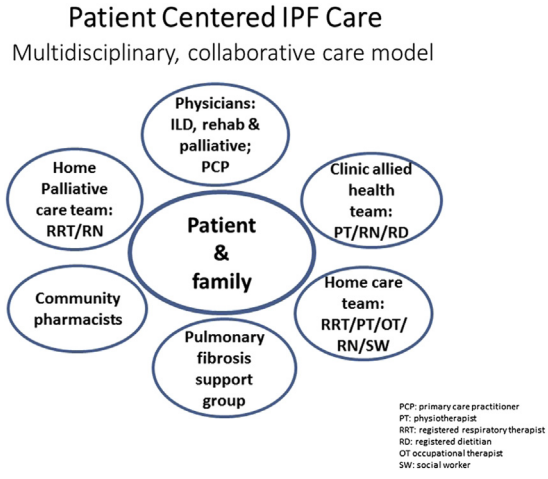

Fig. 1. Multidisciplinary, collaborative care model for IPF. IPF = idiopathic pulmonary fibrosis.

urgent need in IPF care. This philosophy of care negates a demand for a separate needs assessment tool and for identifying a specific time point for referral to a palliative care consultant. We believe that an integrated palliative approach is critical to patient-centered care in IPF.

The second component to achieving a home death is comprised of a multidisciplinary clinic and community team to address the issues identified by an early and integrated palliative care approach. Traditionally, the term "multidisciplinary" in ILD clinics typically refers to "multidisciplinary discussion" to achieve diagnosis by including an expert radiologist and pathologist as emphasized in the guidelines.[4] We strongly believe that a separate multidisciplinary team is essential to ongoing management of an IPF patient, as proposed in the literature (Fig. 1).[16] Our multidisciplinary clinic team consists of an ILD specialist, pulmonary rehabilitation and palliative respiratory care specialist, nurse, physiotherapist, and dietitian. The clinic team collaborates with a community team consisting of the patient's primary care practitioner, home care RRT, and other home care allied professionals are recruited as needed along with a vendor RRT.

The third component for achieving a home death is this close collaboration between community and ILD clinic team. This collaboration between clinic and community teams allows us to provide timely, rapid assessments in the home, enabling efficient modification of symptom management strategies and provide 24/7 support to the patient and his family. This communication strategy allowed the patient to stay out of hospital in the last two months of his life. The organizational structure, funding, and administration of clinic and community teams are distinct, but the teams are seamless in their functioning with respect to patient care. This is engendered by a culture of collaboration and mutual respect with a common goal to support patients and their families in the community. Three way cell phone communications between all parties allows for rapid problem solving and de-escalation of dyspnea crises.

For home deaths to occur, informal caregivers need to be willing, able, and supportive of the patient. We believe that caregiver comfort is the fourth most important component of success in this case report. We have developed dyspnea management strategies that are simple for the caregiver to administer, rapidly effective, and do not require needles. We clearly define the support pathways for caregivers so that they know who to call at any time to help problem solve a concern or crisis. When symptom self-management strategies and health professionals are readily accessible, the caregivers are confident, empowered, and feel supported in their ability to care for their loved ones at home. Our patient's wife in this case was initially reluctant to support a home death. But she changed her mind on experiencing the efficacy of our crisis breathlessness strategies and realizing that she could access support 24/7.

The respiratory literature suggests that breathlessness cannot be managed well at home, and therefore, hospital admission will likely be required at the end of life for symptom control.[10] We, therefore, developed needle-free self-management strategies that can provide rapid and effective symptom relief at home, and this is the fifth important component of our care model to support a home death. It is the bedrock of our approach to early and integrated palliative care. Literature review also suggests that rapid and effective dyspnea management is required to facilitate comfortable home deaths.[17] In our case, effective breathlessness self-management strategies lead to good symptom control, allowing the patient to be engaged in living and enjoying life; our patient remained active until the very moment of his death. In our experience, we are able to achieve good symptom control without compromising on alertness and suppressing intrinsic respiratory drive even in patients with end-stage lung disease. As a result, patients are not only less breathless while engaged in activity but also alert enough to have meaningful conversations with family and friends. Terminal sedation was not required in our patient's case.

Our case study demonstrates that a good home death in IPF can be achieved by early integrated palliative care with collaboration between an academic subspecialty ILD clinic and a community-based home care team. It is possible to attain good symptom control without hospitalization. This model of early and integrated palliative care can provide an acceptable quality of life, allow patients to remain active, and avoid a bedbound state at the end of life in IPF.

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