Holistic management of patients with progressive pulmonary fibrosis

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Shareable abstract (@ERSpublications)
Patients with progressive PF need personalised care plans. Digital healthcare and pulmonary rehabilitation can provide support at every stage. Research is needed to solve technical issues with oxygen supplementation and improve patients' quality of life. https://bit.ly/44KnRc0

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Abstract

Progressive pulmonary fibrosis (PF) is a complex interstitial lung disease that impacts substantially on patients' daily lives, requiring personalised and integrated care. We summarise the main needs of patients with PF and their caregivers, and suggest a supportive care approach. Individualised care, education, emotional and psychological support, specialised treatments, and better access to information and resources are necessary. Management should start at diagnosis, be tailored to the patient's needs, and consider end-of-life care. Pharmacological and non-pharmacological interventions should be individualised, including oxygen therapy and pulmonary rehabilitation, with digital healthcare utilised as appropriate. Further research is needed to address technical issues related to oxygen delivery and digital healthcare.

Educational aims

- To identify the main needs of patients with PF and their caregivers.
- To describe the components of a comprehensive approach to a supportive care programme for patients with PF.
- To identify further areas of research to address technical issues related to the management of patients with PF.

Introduction

Several interstitial lung diseases (ILDs), including idiopathic pulmonary fibrosis (IPF), are characterised by progressive fibrosis (figure 1), and share similar disease behaviour, impacts and needs. These diseases lead

to limitations in daily activities and social participation due to symptoms such as breathlessness, cough and fatigue. In the past decade, significant improvements have been made in pharmacological therapies, mainly for IPF, which have improved prognosis. And while they show little benefit in managing patients' symptoms) [1], they provide a window of opportunity for developing effective practices aimed at relieving symptoms and improving health-related quality of life (HRQoL).

Another important landmark in the evolution of care in those with pulmonary fibrosis (PF) was the coronavirus disease 2019 (COVID-19) pandemic, as it has profoundly changed healthcare services and care delivery policies, particularly the access to non-pharmacological interventions addressing rare pulmonary diseases. Moreover, the pandemic has accelerated access to telemedicine, and we are now living in a hybrid model of providing supportive care. Thus, to better adjust to this new era of healthcare, a comprehensive analysis of the patient's needs, considering which of these needs are unmet, and care opportunities is needed to allow for the proposal of a holistic approach to bundle the different non-pharmacological interventions and ensure continuity of care from early supportive measures to the end of life [2].

In this review, we summarise the main overarching needs of patients with PF and their caregivers. We also describe a holistic management approach addressing the main components of a supportive care programme for these patients. This approach was outlined during a dedicated symposium at the European Respiratory Society International Congress in Barcelona in September 2022 [3] and integrated with a narrative literature review. The different facets of this holistic management programme are presented in four main sections:

1) Patients' (unmet) needs
2) Symptom relief, supportive and end-of-life care
3) Standards of pulmonary rehabilitation (PR) in PF
4) Digital management and organisational aspects

Patients' (unmet) needs

Needs are highly individual and vary throughout the disease course. They include timely and accurate diagnosis, access to pharmacological and non-pharmacological interventions (*e.g.* PR, nutritional and

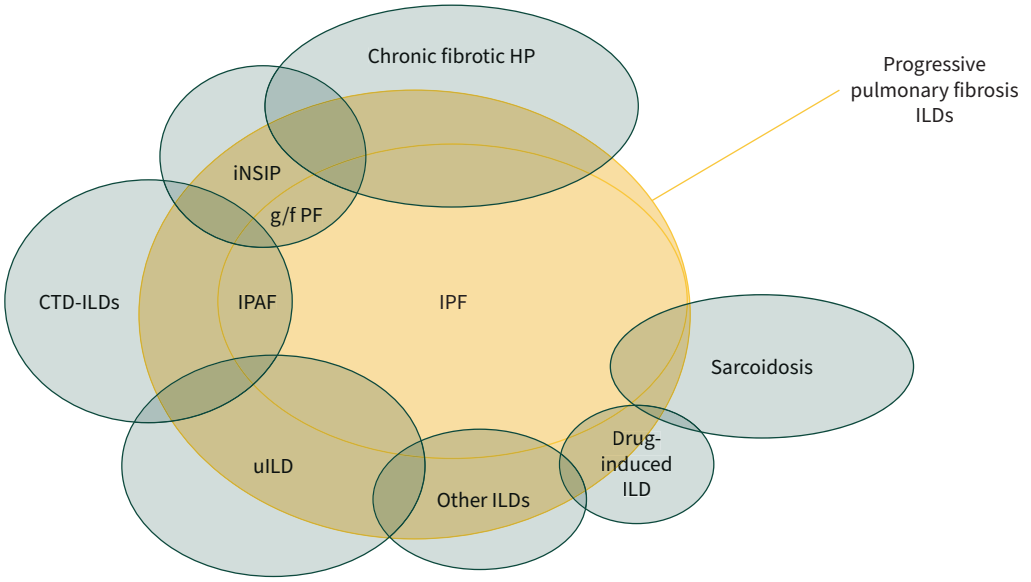

FIGURE 1 Illustration of the different types of interstitial lung disease (ILD) associated with progressive pulmonary fibrosis. ILDs can be broadly categorised into connective tissue disease (CTD)-ILDs, other ILDs, and unclassifiable ILDs (uILDs). The CTD-ILDs are further sub-categorised based on the type of CTD, including rheumatoid arthritis-associated ILD, systemic sclerosis-associated ILD, mixed CTD-associated ILD, and other autoimmune ILDs. Other ILDs include exposure related ILDs (*e.g.* asbestosis and silicosis), non-idiopathic pulmonary fibrosis (such as genetic and/or familial pulmonary fibrosis (g/f PF)), idiopathic interstitial pneumonias (such as desquamative interstitial pneumonia), and others. HP: hypersensitivity pneumonitis; iNSIP: idiopathic nonspecific interstitial pneumonia; IPAF: interstitial pneumonia with autoimmune features; IPF: idiopathic pulmonary fibrosis.

psychological support, oxygen therapy), disease-related education and awareness, and the humanistic burden of ILD, aimed at alleviating symptoms and reducing the impact of PF on patients, families and caregivers [4]. Comprehensive care designed to meet these needs should focus on collaboration and trust among patients, families and the healthcare team.

Diagnostic challenges

The diagnosis of ILD presents significant barriers in providing accurate and timely care. ILD encompasses a diverse group of lung disorders with overlapping clinical manifestations and radiological patterns, leading to complex and challenging diagnostic processes even for experienced physicians [5]. Current diagnostic guidelines define specific radiological and histopathological features for diagnosing IPF [6], but scans and biopsies often show mixed or discordant patterns, making differentiation between IPF and other ILDs difficult [7]. Early diagnosis is also hampered by the subtle and nonspecific nature of initial symptoms, which patients often attribute to ageing or deconditioning, resulting in delayed medical attention [5]. Patient-related risk factors such as male sex, increased body mass index, and gastro-oesophageal reflux can further contribute to diagnostic delays [8, 9]. The involvement of a multidisciplinary team consisting of experienced ILD experts from various specialties becomes crucial in achieving accurate diagnoses. These experts, including pulmonologists, radiologists, pathologists and rheumatologists, enable a comprehensive evaluation of each patient's clinical, radiological and pathophysiological presentation, leading to a more precise differential diagnosis [5]. This collaborative approach not only enhances diagnostic accuracy but also enables the development of individualised treatment plans tailored to the specific ILD subtype of each patient.

However, accessing a multidisciplinary team with experienced ILD experts can be challenging. Limited availability of ILD specialists, especially in certain geographic regions or healthcare systems, may result in delays in obtaining a proper diagnosis [5]. Digital collaboration has emerged as a promising solution. The 2019 international study, STARLINER [10], used a novel digital platform where patients, local hospitals and tertiary ILD referral centres could upload patient's home spirometry, accelerometry, and questionnaires, as well as hospital-collected data like imaging. This facilitated digital multidisciplinary discussions and improved access to expert care. Future research should focus on further optimising the usability and user experience of digital collaboration platforms to ensure seamless integration into clinical workflows and enhance the engagement of healthcare providers (HCPs) and patients.

Pharmacological treatment

Patients with ILD often face significant challenges in accessing pharmacological treatment. One of the primary obstacles is the limited availability and high cost of approved medications for various ILD subtypes [4]. Clear treatment guidelines for specific ILD subtypes, such as hypersensitivity pneumonitis and unclassifiable ILD, are often lacking and heavily reliant on expert opinions [11], further complicating treatment decisions and insurance reimbursement. Geographic disparities and differences in healthcare systems can also affect access to specialised ILD clinics and experts who can prescribe and monitor these treatments [12]. As a result, many ILD patients may experience delays in obtaining appropriate pharmacological therapies, leading to potential disease progression and diminished quality of life. Addressing these challenges requires collaborative efforts among healthcare professionals, regulatory authorities, and patient advocacy groups to advocate for improved access to effective pharmacological treatments for ILD patients.

Pulmonary rehabilitation

A European Pulmonary Fibrosis Federation (EU-PFF) survey, conducted in approximately 18 European Union countries with patients and HCPs about their perspectives on the care received by patients with PF, highlighted problems with self-management, and with patients feeling they had insufficient information and support [13]. Despite established scientific evidence and patients' experience of the benefits of PR to their overall health and mental health, in half of the countries surveyed patients do not have access to PR, including some patients in high-income countries like Spain and UK [13]. Even when PR is available, it may not offer education specialised to the needs of patients with PF. Most PR programmes tend to focus on providing general education about respiratory diseases, with a slight tendency towards COPD components, and may not address the specific needs of patients with PF. As a result, patients miss out on the opportunity to connect with others who have similar challenges and learn coping strategies specific to their condition. Furthermore, according to Bonella *et al*. [14] these classes may provide limited support to the patient's carer/family and were often held in locations that were challenging to access *(e.g.* too many stairs) or too far away for a patient with severe breathlessness.

Nutrition

Patients with ILD may be at risk of malnutrition due to the disease process and associated medical treatments [15], resulting in negative impacts on exercise capacity, quality of life, and mortality [16–18]. Research in this patient population is limited, but existing evidence suggests that malnutrition affects approximately 9% to 55% of individuals with ILD, depending on the assessment methods used [17, 19]. While body mass index has been commonly used to assess nutrition status and survival in ILD patients, examining fat-free mass may provide more valuable insights for improving patient outcomes [20]. Additionally, vitamin D plays a crucial role in the lung immunological process, and screening for deficiency and appropriate treatment may be beneficial [20]. However, further research is necessary to establish clear guidelines.

Ongoing nutrition management and counselling, based on evidence-based approaches, are essential to optimising nutrition outcomes for ILD patients. Collaboration and data sharing among healthcare clinicians in academic teaching hospitals can facilitate larger sample sizes, leading to more meaningful conclusions and increased statistical power in research studies related to ILD and nutrition [20].

Counselling/psychological support

Another unmet need is the lack of routine counselling or psychological support services for patients with IPF, and inadequate emotional support for carers and families, particularly leading up to, and following, the death of a patient [21]. Where available, PR was reported to be a source of psychological support to patients with IPF; however, access to PR varied widely between countries. Klein *et al.* [22] underlined lack of access to comprehensive patient-centred care, and the need for a multidisciplinary team to facilitate the diagnosis and management of patients with IPF, including radiologists, ILD specialists and specialist nurses. The need for physiotherapists as part of the multidisciplinary team was also highlighted.

Oxygen therapy

Oxygen supply reimbursement was problematic in a third of the European countries surveyed, which may compromise access for patients with limited resources and generate anxiety [13]. Additionally, oxygen equipment is often heavy and difficult to use, highlighting the need for collaboration between HCPs, technology companies and patients to develop equipment that meets patients' needs. Challenges surrounding oxygen use include inadequate patient training on proper usage, as well as a lack of awareness among HCPs regarding distinguishing oxygen needs between patients with COPD and PF. Virtual care can provide certain advantages, such as the ability for patients to transfer large amounts of health information (*e.g.* symptom scores, vital signs, oxygen saturation levels) to HCPs and the convenience of being able to communicate with HCPs from home. However, patients sometimes feel that the quality of communication and overall care may be reduced during virtual consultations. Thus, timing and opportunities for virtual care should be agreed upon between patients and physicians before its implementation [23].

Disease-related education and awareness

Need for information is one of the main issues for patients with PF. They crave information on several areas of the disease including, but not limited to, the management of cough and breathlessness, treatment options such as PR, transplantation, and end-of-life options [14]. Qualitative studies have shown that patients and carers have distinct information needs at different disease stages and seek information in different formats and from different sources [24, 25]. Specifically, patients report the need to be provided with information about their diagnosis as soon as possible to "know what they were up against" and "how to live their lives to the fullest" despite the consequences of the disease. They want updates at each visit regarding novelties in the field and specific practical guidance on how to travel with oxygen.

Finally, patients want to know about their prognosis but also to hear "success stories" of patients defying prognostic timelines to boost their positivity and hope [25]. In more severe disease, carers may have more need for information about prognosis than the patients [24]. Patients expect this information to be presented in several complementary ways, such as explained verbally at clinic appointments, by telephone, in educational presentations, and *via* support groups, but also written in pamphlets, brochures, books, social media, websites and electronic patient portals. Written information is perceived as a safety net that could be consulted at any time but also shared with loved ones. ILD specialist nurses and patient charities are key sources of information. Patients report seeking information from ILD specialist nurses when needing practical advice and triggering medical attention when required [24]. They also have a special role in linking patients to other HCPs and are involved in a variety of roles including being the main point of contact for patients during the pandemic [26]. Patient charities were central sources of information during the pandemic. Action for Pulmonary Fibrosis (APF), a UK-based patient organisation runs a helpline, which patients can access by phone or e-mail. During the COVID-19 pandemic APF launched a befriender

service for isolated PF patients, which provided peer support by phone with trained volunteers [27]. It is also piloting a scheme where specialist centres can refer patients, at diagnosis, to peer support provided by APF. Besides the role of ILD centres and ILD nurses as sources of information and support, patients with IPF reported that meeting other patients with IPF is very helpful, for example at peer support groups, online forums or when attending PR programmes.

Humanistic burden of ILD

A recently published review focused the humanistic burden of progressive PF and the impact on patients' daily lives, careers and personal relationships [28]. Dyspnoea, cough, and fatigue may lead to low mood and isolation, by limiting the patient's ability to carry out physical tasks and participate in social activities [29–31]. Alterations of emotional and mental wellbeing may occur, with symptoms of stress, anxiety, depression, and in some cases, suicidal thoughts [29–31]. The usual labour activities may be limited as well as the patient's ability to look after themselves, forcing them to rely on caregivers for help [29–31]. A high caregiver burden has been also reported that may strain relationships between caregivers and patients [28].

Symptom relief and supportive care end-of-life care

Respiratory and extrapulmonary symptoms contribute significantly to the impaired HRQoL experienced by patients with PF and their families. Breathlessness, cough, fatigue, anxiety and depression are the most commonly reported symptoms and tend to worsen with disease severity. Symptom management should begin upon diagnosis and be tailored to the patient's needs throughout the disease course, including end-of-life care. Supportive care for patients with PF is multifaceted, involving both pharmacological and non-pharmacological interventions, focusing on both disease-modifying treatments and concurrent supportive measures, such as supplementary oxygen, symptom control strategies, education and end-of-life care [2, 13]. Using a supportive care approach, we will describe various options for managing symptoms, evidence for oxygen supplementation and end-of-life care for patients with PF. Figure 2 summarises the key topics discussed in each area addressed.

Breathlessness

Breathlessness is the most common and often the initial symptom experienced by patients with ILD [1, 32]. The symptom arises from a multitude of factors, but, in brief, reflects an increased ventilatory drive and/or decreased ventilatory capacity. Briefly, the ventilatory drive is generated by the brain's predictive mechanisms of assessing the breathing requirements to supply the body's ventilatory needs at a given time. The prediction is conveyed *via* efferent pathways to the respiratory system, including the airways, lungs and respiratory muscles. Conversely, afferent information is conveyed to the central nervous system from

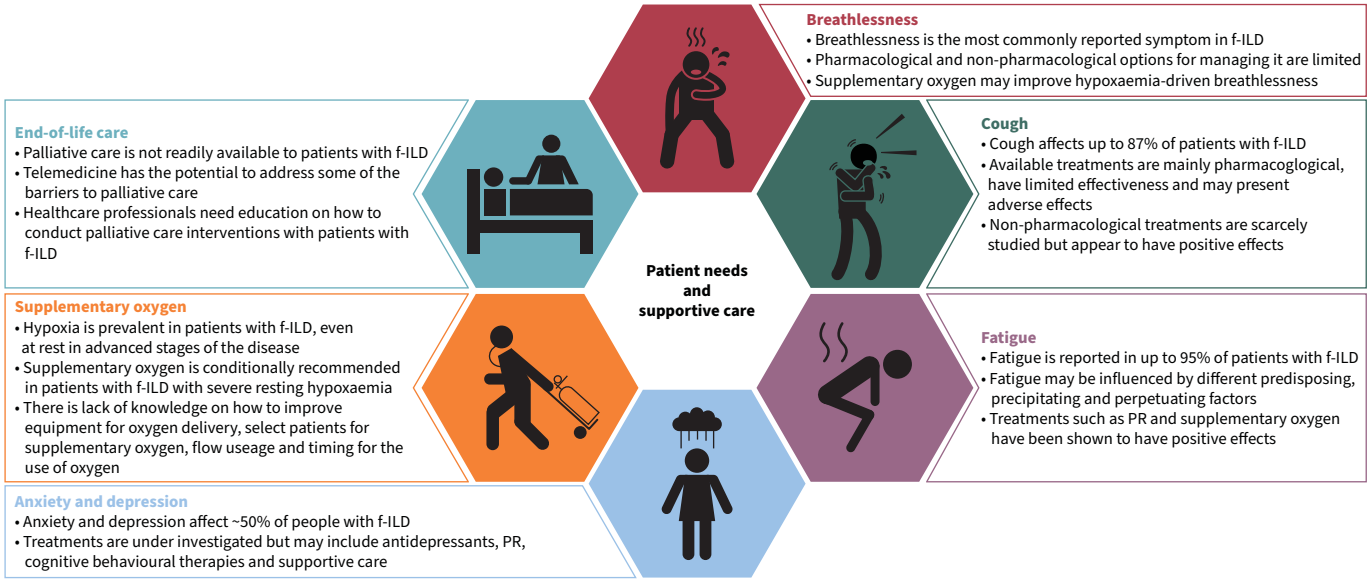

FIGURE 2 Summary of the current knowledge, gaps and opportunities for supportive care in needs identified by patients with pulmonary fibrosis. f-ILD: interstitial lung diseases associated with progressive pulmonary fibrosis; PR: pulmonary rehabilitation.

various systems, such as the musculoskeletal, respiratory and cardiac systems, regarding their needs and supply [33]. If the ventilatory response and work of breathing match the predicted need, the individual remains unaware. However, if there is a mismatch between the drive and capacity to drive or a perceived threat to breathing, breathlessness is triggered and perceived by the person. Several factors may contribute to this mismatch, such as hypoxaemia (*i.e.* low levels of oxygen in the blood), respiratory mechanical constraints, muscle dysfunction and fatigue, anxiety, loss of control, and influence of other symptoms.

Although breathlessness is strongly associated with poor HRQoL [34], options for managing the symptom are limited. Antifibrotic drugs have not been shown to affect breathlessness, benzodiazepines lack evidence but may relieve acute anxiety, and the evidence base for opioids is limited, with no consistent evidence for efficacy in outpatients with respiratory disease [35–37]. Moreover, in clinical practice, their use may be limited due to safety concerns. Oxygen supplementation has been suggested as a potential treatment for reducing breathlessness levels during exertion [38]. Such treatment could be potentially beneficial for patients in whom the main drive for breathlessness is hypoxaemia, which is present during exertion at diagnosis in ∼40% of patients with IPF and 70% after 5 years [39]. In an anaerobic environment, the peripheral skeletal muscles produce lactic acid, and ventilation increases sharply to buffer this. Administering oxygen can delay this mechanism, decrease the breathing effort, and prolong the time the person can exercise without having to stop due to distressing breathlessness (see figure 3). However, there is still a need for evidence supporting the benefits of oxygen supplementation in reducing dyspnoea during daily activities before it can be widely recommended.

Cough

Cough affects up to 87% of patients with PF [40] and is responsible for major physical and mental impairment, has associations with respiratory hospitalisation, death and lung transplantation [41], and increases societal costs (productivity loss costs EUR 7952 year per employee) [42]. The burden of cough, specifically chronic cough (*i.e.* a cough that lasts for more than 8 weeks), has led international guidelines to highlight it as a key symptom to address [43]. However, treating chronic cough is challenging as there is currently no licensed treatment for this symptom in ILD. Available treatments are mainly pharmacological, have limited effectiveness [44, 45] and significant adverse effects, such as drowsiness and risk of addiction [46]. There is a paucity of evidence on non-pharmacological interventions in patients with chronic respiratory diseases, specifically in PF [47]. Observational and case studies have shown that multicomponent programmes consisting of education, vocal/laryngeal hygiene, hydration, cough control techniques, and psychoeducational counselling can help. These programmes can increase a patient's cough-related quality of life and empower them to take control over their cough [48].

Fatigue

Fatigue is reported in up to 95% of patients with PF, a much higher prevalence than in the general population (5–20%) [49]. The aetiology of fatigue in ILD is multifactorial and may be influenced by different factors predisposing (*e.g.* biological and personality), precipitating (*e.g.* psychological and social stress) or perpetuating (*e.g.* other symptoms, and poor coping and social support) the fatigue. However, many

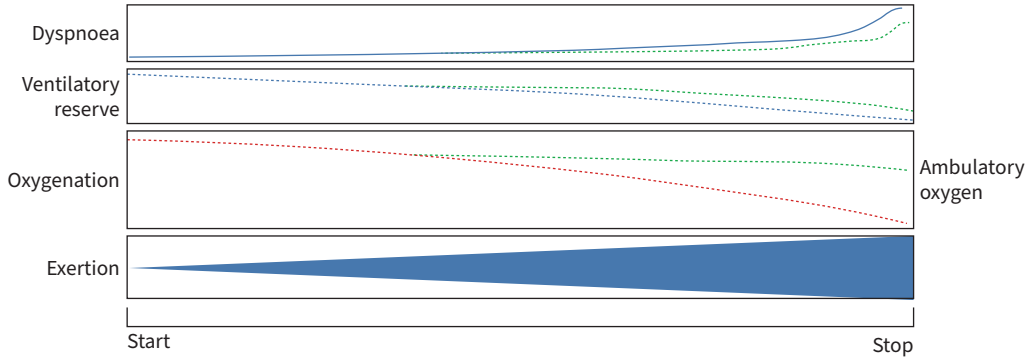

FIGURE 3 Illustration of potential effects of supplemental oxygen in a patient with pulmonary fibrosis and hypoxaemia during exertion. Green lines represent potential responses with supplemental oxygen, and blue and red lines the potential responses without supplemental oxygen. Responses are not observed, the data for illustrational purposes only. Adapted from the presentation given by Magnus Ekström (Lund, Sweden) at the 2022 European Respiratory Society International Congress, with permission from the author.

mechanisms are still unknown [49]. Fatigue is a significant burden for patients with ILD and can negatively impact social relationships, work participation and HRQoL. It is often misunderstood by family, friends, employers and HCPs, exacerbating social isolation. Fatigue is associated with depression, anxiety, decreased work participation and loss of income, and is a predictor of a lower HRQoL in patients with ILD [49]. Guidelines and evidence-based treatment recommendations for fatigue are still lacking, but treatments such as PR and supplementary oxygen have shown positive effects in reducing tiredness and exhaustion [2]. To optimise the treatment of fatigue in ILDs, a personalised, multidisciplinary approach seems to be the most suitable. Such an approach should be comprehensive and tailored to each individual's unique needs.

Anxiety and depression

Depression and anxiety are more common in patients with PF than in the general population. It is estimated that depression affects 14–49% and anxiety 21–60% of patients with PF [50]. Anxiety and depression are associated with adverse consequences by impairing HRQoL, increasing hospital admissions and length of hospital stays, increasing exacerbations, leading to poor adherence to medical treatment and dependency on caregivers, reducing social interactions, and contributing to premature death [50].

Although highly prevalent and a significant burden on patients, anxiety and depression are frequently overlooked and often undertreated or even untreated [34]. Interventions may include antidepressant drug therapy, PR, cognitive behavioural therapies and supportive care [51].

Oxygen supplementation

Hypoxia is prevalent in patients with PF, even at rest, in advanced stages of the disease. Nevertheless, evidence on the effects of supplemental oxygen for patients with PF is scarce. Two major guidelines exist, which conditionally recommend the use of supplemental oxygen in patients with PF with severe resting hypoxaemia [52, 53]. However, the indications are not precise and are based on data from studies on patients with COPD, who often are different in terms of mechanisms of exercise limitation and hypoxaemia compared with patients with PF [54]. The primary goal of using supplemental oxygen in patients with PF is to protect organ function which can potentially enhance physical functioning, improve HRQoL and survival. Nonetheless, there is a paucity of data to guide the appropriate use of supplemental oxygen in PF, in terms of criteria for the selection of patients, equipment, flow rates and timing for the use of oxygen.

Evidence exists regarding the efficacy of supplemental oxygen during standardised exertion testing. However, it is uncertain whether such benefits translate to improvements in day-to-day physical functioning or HRQoL [55]. The AmbOx trial, the first randomised controlled trial of its kind, demonstrated that patients with isolated exertional hypoxia (peripheral oxygen saturation $\leqslant$88% on a 6-min walk test (6MWT) due to PF who used lightweight ($\sim$2 kg) ambulatory oxygen cylinders during daily activities for 2 weeks had an improvement in day-to-day physical functioning and HRQoL. At the end of the trial, two-thirds of patients chose to continue using ambulatory oxygen [56]. However, it should be noted that this was a non-blinded trial and only a 2-week treatment period was performed.

Due to the practical and psychological challenges associated with using oxygen, as well as the limitations of current ambulatory oxygen delivery devices, a trial of supplemental oxygen designed for and with patients with PF and exertional hypoxia should be considered, to facilitate an informed, shared decision. HCPs must provide proper education, follow-up and monitoring for side-effects to ensure the treatment's safety, efficacy and proper use.

Nocturnal hypoxaemia is a common occurrence in PF, even in patients without daytime resting hypoxaemia, and is often associated with pulmonary hypertension and poor prognosis. Administering supplemental nocturnal oxygen to correct nocturnal hypoxaemia may prevent or slow down the development of pulmonary hypertension, although there remains a paucity of data addressing this possibility in PF. Some guidelines recommend nocturnal oxygen for patients who desaturate below 88% for at least one-third of their total sleep time [57, 58]; however, no evidence of benefit was observed for people with COPD [59]. Studies conducted in pulmonary hypertension suggest administering nocturnal oxygen to patients who experience desaturation <90% for more than 10% of their total sleep time [60]. Data are needed to guide prescription of nocturnal oxygen in patients with PF without daytime hypoxaemia, including the threshold of desaturation and duration of desaturation events which should trigger use of supplemental oxygen.

End-of-life care

Despite being recognised as a necessity by patients, caregivers and HCPs, end-of-life care is not readily available to patients with PF. According to Harrison *et al.* [61], only one-third of patients are referred to palliative care when they initiate long-term oxygen therapy, even though the mean survival time at this

stage is only 1.5 years. The barriers to referral include a perceived lack of local access to palliative care, lack of established optimal timing for referral, insufficiently specialised professionals, insufficient knowledge of the specific palliative care needs of patients with PF, and a lack of collaboration between palliative care and respiratory teams, as well as difficulties travelling, transportation difficulties, severe breathlessness and high supplemental oxygen requirements among patients [13, 62]. Digital health shows potential to address these inequalities in health access. It's benefits and drawbacks will be discussed in detail in the section focusing on "Digital management and organisational aspects".

A World café qualitative study with patients, caregivers, HCPs, industry representatives and a member of the clergy addressed six topics on palliative care, including legal matters and end-of-life conversations, preferred place of dying and treatment limitations. The group highlighted the role that advocacy groups for patients can have to push for PF to be recognised as a critical illness for insurance purposes and acknowledged how difficult end-of-life conversations are. They concluded there was a need to study and address HCPs needs regarding end-of-life conversations and currently the Irish Lung Fibrosis Association is developing a conversation guide on this matter [63].

Standards of PR in PF

Rehabilitation in ILD: new evidence for benefit in PF

Evidence of the effects of PR for patients with PF has increased since the first Cochrane review in 2008, which included five randomised controlled trials (RCTs), to the most recent review published in 2021 with 21 studies [64]. The 2021 Cochrane review confirms the short-term benefits of PR and adds new information suggesting that there may be sustained benefits of PR after 6–12 months for physical function, breathlessness and HRQoL. The sustained differences between groups in the long term seem to be more related to significant declines in participants from the control groups [64]. There is also emergent evidence that PR completion is associated with a survival benefit in PF. Data from 701 patients collected from PR programmes around the world show that completing more than 80% of sessions is associated with a 30% survival increase. The results are particularly evident in participants starting PR with low functional capacity (6-min walking distance <350 m) and improving above the minimum clinically important difference [65].

Rehabilitation in ILDs: referral and uptake

Several international respiratory societies have been recommending PR as the standard of care for patients with PF since 2011; however, referral and uptake are still low. National registry data from Germany (Insights-IPF 2015), Spain (SEPAR National Registry 2019) and the USA (IPF-PRO 2021) indicate that <20% of patients (19.5% in the USA) are referred for PR and <10% attend (4.5% in Germany and 10.1% in Spain) [66–68]. Barriers to PR uptake in PF are very similar to those reported in other patient groups: travelling and transportation, having to manoeuvre oxygen tanks, competing demands including caring responsibilities, the need to take time off of work, side-effects of medications, and feeling that the burden of PR outweighs its potential benefits [69].

Home-based and telerehabilitation

The delivery of PR has been changing over the years to overcome barriers to its access and motivated by the emergent technology that allows remote supervision and monitoring. The COVID-19 pandemic has accelerated the uptake of home-based and remote PR programmes, although the sustainability of such models beyond the pandemic is unclear. As shown in the UK National Asthma and COPD Audit Programme 2021, home PR moved from being virtually non-existent in 2015 (n=1) to being offered by 86.5% of PR services in 2021 [70]. There is emerging evidence for the benefits of telerehabilitation in chronic respiratory diseases, although at this stage data are scarce in ILD. A recent Cochrane systematic review has shown similar short-term effects of telerehabilitation in comparison with centre-based PR for physical function in patients with chronic respiratory diseases [71]. For ILD, there is evidence from a programme evaluation [72] and two pilot RCTs [73, 74] that telerehabilitation is safe and results in clinically meaningful gains in HRQoL, 6MWT and physical activity compared with centre-based PR or usual care. However, the composition of the interventions were heterogeneous, including real-time supervision programmes, bespoke platforms and remote monitoring apps. Two-thirds of studies did not have a sample size calculation and one was unable to demonstrate equivalence to centre-based PR for HRQoL. Real-world experience from the Melbourne centre indicates that although telerehabilitation may be adequate for the majority of patients with PF, around 20% of them are unsuitable due to fall risk, high oxygen requirements on exercise, the need for additional monitoring (*e.g.* pulmonary hypertension), cognitive deficits, communication difficulties, an unsuitable home environment and preferences for centre-based PR.

In conclusion, there is increasing evidence for the benefits of PR possibly sustained to 12 months and its delivery has been improved through the addition of home-based and remote PR programmes with emerging evidence in PF. Future challenges are optimal patient selection for the different PR settings.

Digital management and organisational aspects

During the COVID-19 pandemic, digital management has gained popularity for patients with PF, providing several benefits such as monitoring disease progression, guiding treatment decisions, and improving self-management and non-pharmacological interventions. Digital management tools, including wearable devices, telemedicine platforms and mobile apps, can help patients monitor their physical activity, track symptoms and medication adherence, and communicate with HCPs. This can lead to early interventions and improved disease management. Digital management can also simplify clinical research by facilitating direct communication between patients and practitioners, improving treatment adherence, monitoring side-effects, and enabling international real-world registries [75].

Current knowledge about eHealth in PF

In the last decade, the definition of "health" has shifted towards supported self-management due to an ageing population and increased demand for healthcare services, which is difficult to sustain in developed countries. Simultaneously, digital health has gained acceptance among HCPs and patients, especially during COVID-19. A recent survey of 286 specialists in ILDs from 54 countries revealed that ∼40% used eHealth tools, such as home-based oxygen saturation, symptoms and HRQoL scores, and home spirometry. Positive experiences were reported by 96.5% of respondents, primarily for improving quality of care [76]. Patient satisfaction with home monitoring was high in a single centre study with 166 patients with PF, and the healthcare institution was able to replace 50% of their outpatient visits with remote consultations [77].

In patients with ILD, hybrid models of care including face-to-face and digital support may offer an opportunity to reduce the personal burden of travelling to specialist centres and poor access to specialist care by supporting patients at home throughout the disease course. Hybrid models can: support the peri-diagnostic period to follow-up patients at risk of developing ILD (*e.g.* scleroderma), facilitate early diagnosis and guide treatment decisions; follow-up for early detection of disease progression and/or acute exacerbations (including post-transplantation); support disease management by immediate monitoring and management of side-effects, implementation of telemedicine and telerehabilitation, or local rehabilitation with remote specialist information; and facilitate end-of-life care to help reach HCPs in a phase when it is difficult to travel or move out of the home. In the future, digital tools could even ease cross-border care, enabling patients to access foreign experts.

Spirometry is one of the outcome measures showing most potential for use in the follow-up and monitoring of PF. Home monitoring of lung function could help identify exacerbations [78], provide early predictions of disease progression [79] and detect treatment effects, allowing early dose tailoring and prevention of side-effects [80]. This topic will be assessed in a follow-up study of the Dutch national trial PREDMETH [81]. However, further verification of its validity and reliability is needed. Single centre studies have shown that data acquired from home is reliable when compared with hospital spirometry [82–84]. However, multicentre studies have reported problems with implausible outliers, long-term missing data, and weak correlations between longitudinal patterns in home and hospital spirometry [85, 86]. Artificial intelligence may help in developing algorithms to standardise acceptability and usability of the spirometric manoeuvre and deliver real-time automated feedback to patients [87].

Other increasingly used devices include pulse oximetry, activity trackers and apps collecting patient-reported outcome measures (PROMs). A prospective single centre study in Germany with 46 IPF patients found that the average number of steps per day was a better predictor of mortality than the Gender–Age–Physiology (GAP) Index, although day-to-day variation needs to be considered [88]. PROMs can be administered mainly through the use of digital visual analogue scales and physical function through the use of simple surrogate measures, such as sit-to-stand tests and collaborations with known technology developers (*e.g.* the Stanford-Apple collaboration 6MWT at home) [89, 90]. Additionally, there is qualitative evidence that home monitoring tends to clinically improve the psychological well-being of patients with PF [91], with patients reporting feeling "more in control, that they had more tools to manage their disease".

Telemedicine for end-of-life care has been implemented by Bischoff *et al.* [92] in a post-COVID-19 ILD pilot programme in which nearly half of the visits were conducted *via* videoconferencing. At baseline, less than half of the patients felt that they had received sufficient information about the disease and treatment options but, after an average of three visits, patients and their families reported improved symptom

management and a better understanding of the illness. Additional benefits included the convenience of not having to travel to the palliative care centre and the possibility of having several family members participate in video consultations. This study highlights that HCPs need to think strategically about which patients to refer to palliative care, using telemedicine to increase access for symptomatic patients on high levels of oxygen who live far away from the centre; developing patient-facing materials and scripts for introducing palliative care to patients, and training HCPs on symptom management; and advanced care planning conversations in centres that do not offer palliative care [92].

Challenges for implementation and upscaling

The development and implementation of any innovative idea is challenging. Regarding home spirometry several questions are pending, such as determining the optimal frequency of measurements, standardising initial patient training, and the possibility of using automated alerts for patients and physicians. Linear regression models are being investigated to predict longitudinal lung function depending on the number of data points and observation time [86]. Furthermore, the ability of home spirometry to detect acute exacerbations of IPF is currently being studied in a German prospective multicentre trial (ClinicalTrials.gov identifier: NCT03979430).

Interventions with multiple devices must be evaluated during clinical trials to validate their feasibility and their potential in therapeutic management of ILDs. A number of trials are currently underway, including an observational international multicentre study monitoring 700 patients with PF using home spirometry, pulse oximetry and PROMs sent directly to the hospital [93]. Another prospective randomised study is designed to compare the standard of care with a telemonitoring programme in patients with connective tissue disease-associated ILD. Telemonitoring will consist of spirometry, oximetry, pulse and blood pressure measurement, activity measurement and questionnaires about cough and breathlessness. These trials aim to provide insights into the benefit and feasibility of digitally supported tools in the management of ILDs [94]. These trials will give critical insight into the feasibility and benefit of these digitally supported tools.

Digital healthcare implementation should also consider the possible inequalities in access that may arise. Research shows that telemedicine encounters may hinder Black patients of lower socioeconomic status from accessing care. Practices with many elderly patients may limit access if the platform is not user-friendly and may benefit from additional technical support. Practices predominantly serving those with lower education and lower socioeconomic backgrounds, including structural reimbursement, may need to consider effective ways to convey medical information to maximise understanding and outreach for all [95]. Other ethical implications include digital literacy, responsibilities in the case of disease progression, and changes in doctor–patient relationships. Wearable devices that collect reliable data passively should be developed and incorporated into electronic patient records for appropriate sharing between patients and healthcare settings. Data storage must comply with privacy and confidentiality regulations and data ownership must be clarified. The ERS Clinical Research Collaboration CONNECT will explore many of these questions to facilitate innovative digital healthcare implementation [96].

Finally, in addition to improving the efficiency of digital tools with the use of AI algorithms, the patient's long-term outcome, safety and psychological well-being have to be closely assessed when replacing hospital care with telemedicine or implementing new monitoring. For example, the potential consequences for patients of viewing their real-time data, the possible delay in medical contact due to a false sense of security, or incorrect medical decision-making due to inappropriate results. All these possible caveats must be followed during a prospective study to allow wider use of these new innovative tools.

Conclusion

Patients with PF have unique needs that require a comprehensive care plan tailored to their individual requirements. These needs include timely and accurate diagnosis, access to treatment, education, emotional support, and non-pharmacological interventions. However, patients with PF struggle with self-management due to a lack of information and support. Support should begin at diagnosis and be customised to the patient's needs, including end-of-life care. PR has demonstrated sustained benefits beyond 6 months, but referral and uptake are still low, probably due to barriers such as transportation, debilitating symptoms and competing demands.

Digital healthcare has emerged as a potential solution to support people with PF to live with their condition. It can offer patients support at home throughout the disease course, from the peri-diagnostic period to follow-up, early diagnosis, treatment decisions, disease management, and palliative care. Further research is required to address technical issues related to digital healthcare. Nevertheless, the use of

artificial intelligence has the potential to standardise spirometric manoeuvre acceptability and usability, improve algorithms, and provide real-time automated feedback to patients.

Key-points

- Management of PF should begin at diagnosis, be tailored to the patient's needs, and consider end-of-life care.
- Patients' needs include individualised care, specialised treatments, education, emotional and psychological support, and better access to information and resources.
- Pharmacological and non-pharmacological interventions should be planned to target individual complaints, including oxygen therapy and PR, and digital healthcare should be used as appropriate.
- Research is needed to address technical issues related to oxygen delivery and digital healthcare in managing PF.

Self-evaluation questions

1. What are the unmet needs of patients with PF?
    a) Timely and accurate diagnosis.
    b) Emotional support and non-pharmacological interventions.
    c) Routine counselling/psychological support services.
    d) All of the above.
2. Which of the following is not a commonly reported symptom experienced by patients with PF?
    a) Breathlessness.
    b) Memory problems.
    c) Cough.
    d) Fatigue.
3. Considering PR for patients with ILD, which of the following is true?
    a) Rehabilitation has no sustained benefits after 6–12 months for physical function and breathlessness.
    b) Telerehabilitation does not improve physical function in ILD.
    c) Less than 20% of patients are referred for PR and attend the programme.
    d) Telerehabilitation is not suitable for the majority of patients.
4. Which of the following statements is true regarding home monitoring in patients with ILD?
    a) Home monitoring is only useful for patients with scleroderma who are at risk of developing ILD.
    b) Digital healthcare in ILD patients could support them at home throughout the disease course and enable patients to access foreign experts.
    c) Home monitoring has not been well accepted by healthcare providers and patients.
    d) Spirometry is the only outcome measure that has been studied for its measurement properties and practicability to be used remotely in patients with PF.

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Suggested answers

1. d.
2. b.
3. c.
4. b.