Journal of Physiotherapy 68 (2022) 158–164

Journal of PHYSIOTHERAPY

journal homepage: www.elsevier.com/locate/jphys

Research

Physiotherapy management of interstitial lung disease

Anne E Holland

Departments of Physiotherapy and Respiratory Medicine, Alfred Health, Melbourne, Australia; Central Clinical School, Monash University, Melbourne, Australia; Institute for Breathing and Sleep, Melbourne, Australia

KEY WORDS

Interstitial lung disease
Physical therapy
Restrictive lung disease
Pulmonary rehabilitation

Introduction

The interstitial lung diseases (ILDs) are a group of over 200 chronic lung conditions characterised by dyspnoea on exertion, troublesome cough, exercise intolerance, and poor health-related quality of life. Although ILDs vary in underlying diagnoses and clinical course, comprehensive supportive care is considered critical across all ILD subtypes to optimise clinical outcomes and patient wellbeing. Key elements of supportive care for ILD include pulmonary rehabilitation, supplemental oxygen, education, psychosocial support, symptom management, and end-of life care (Figure 1).[1] This review summarises: the classification and causes of ILD; the burden of this condition for individuals and the health system; the clinical features of ILD; the key elements of comprehensive ILD care focusing on interventions delivered by physiotherapists; and future directions for research and practice.

What is interstitial lung disease?

The ILDs are a group of restrictive lung conditions characterised by inflammation and/or fibrosis of the lung tissue. In most of these conditions the pathologic abnormalities occur predominantly in the lung interstitium, which is the connective tissue framework surrounding the alveoli, airways and blood vessels. The ILDs can be grouped into five broad clinical categories (Figure 2), depending on whether there is a known cause such as an underlying disease process; exposure to environmental toxins; exposure to radiation or drugs known to cause ILD; presence of a connective tissue disease; or if the cause is unknown.[2] Recently, development of pulmonary fibrosis after coronavirus 2019 (COVID-19) infection has been reported, most commonly in those who have undergone mechanical ventilation.[3] Persistent radiographic changes in the lungs have also been demonstrated in survivors of previous coronavirus pandemics, the severe acute respiratory distress syndrome (SARS) and Middle East respiratory syndrome (MERS), although these were generally mild and stable over time.[4] Little is known about the clinical course of pulmonary fibrosis in COVID-19 survivors.

Burden of interstitial lung disease

Although each of the ILDs is relatively rare, collectively the ILDs affect a large number of people across the world. The Global Burden of Disease Study estimated the prevalence of ILD at 82/100,000 people.[5] The most common fibrotic ILD is idiopathic pulmonary fibrosis (IPF), which is a progressive condition that occurs most commonly in men aged > 65 years and confers a poor prognosis. Five-year survival for people with IPF has been estimated at 46% (95% CI 42 to 50),[6] contributing 0.26% of global all-cause mortality.[5] Recently it has been recognised that a substantial proportion of those with non-IPF fibrotic ILDs also exhibit a progressive phenotype known as progressive-fibrosing ILD (PF-ILD) with a similar mortality risk to IPF.[7] Frequent diagnoses in those with PF-ILD include chronic hypersensitivity pneumonitis, autoimmune ILDs, sarcoidosis and idiopathic nonspecific interstitial pneumonia. Other ILDs in which inflammation is more prominent than fibrosis (eg, acute hypersensitivity pneumonitis, cryptogenic organising pneumonia, some connective tissue diseases) have a less progressive course and better survival.[8]

The ILDs are associated with a high symptom burden, including dyspnoea (54 to 98% of patients) and a chronic dry cough (59 to 100% of patients).[9] Fatigue and exhaustion may be more bothersome than dyspnoea for some patients.[10] Impairments in health-related quality of life are substantial, including levels of distress related to breathlessness that are frequently higher than in people with chronic obstructive pulmonary disease (COPD), fatigue worse than in heart failure, depression comparable to people with a major depressive disorder, and sleep disturbance similar to those with obstructive sleep apnoea.[11] The physical, emotional, social and financial burdens experienced by caregivers of people with ILD are increasingly being recognised, particularly as their loved one's disease becomes more severe.[12]

The direct costs of ILD to the health system are substantial and rising. A recent systematic review reported an annual median cost of US$32,384 per patient (range $1,824 to $116,927 per patient), with significantly increased costs since 2014 after new anti-fibrotic

https://doi.org/10.1016/j.jphys.2022.06.006

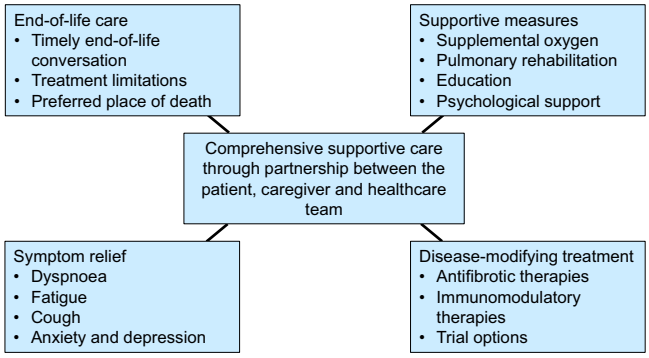

Figure 1. Comprehensive supportive care for patients with interstitial lung disease. Adapted from Wijsenbeek et al.[1]

medications for IPF were introduced into practice.[13] In the USA, annual direct medical costs for patients with IPF were estimated to be twofold higher than age-matched and gender-matched controls in the year following IPF diagnosis.[14] The main contributors to healthcare costs were inpatient (55%), outpatient (22%) and medication costs (18%); costs varied substantially across healthcare systems. There are few data on indirect costs, although work productivity loss in fibrotic ILD has been estimated at US$9,313 to $10,902 per patient per annum.[13] Costs for work productivity loss amongst caregivers are unavailable, but are likely to be significant.

Clinical features of interstitial lung disease

People with ILD generally present with breathlessness and/or cough, often resulting in reduced exercise tolerance. A high-resolution computed tomography of the chest reveals radiological features of ILD, which vary according to the ILD subtype. The diagnosis of an ILD is a complex process that requires integration of clinical, radiological and histopathological data, frequently in a specialist multidisciplinary meeting.[15] Achieving an accurate diagnosis of the ILD subtype is important, as it determines eligibility for specific pharmacotherapies. Respiratory function tests typically reveal reductions in forced vital capacity and diffusing capacity for carbon monoxide. Reduced functional exercise capacity is often demonstrated on 6-minute walk test, and marked exercise-induced desaturation may also be present. Measures of respiratory function and exercise capacity are repeated at regular intervals to track progression of disease over time. Development of hypoxaemia, either at rest or on exertion, confers a poor prognosis.[16]

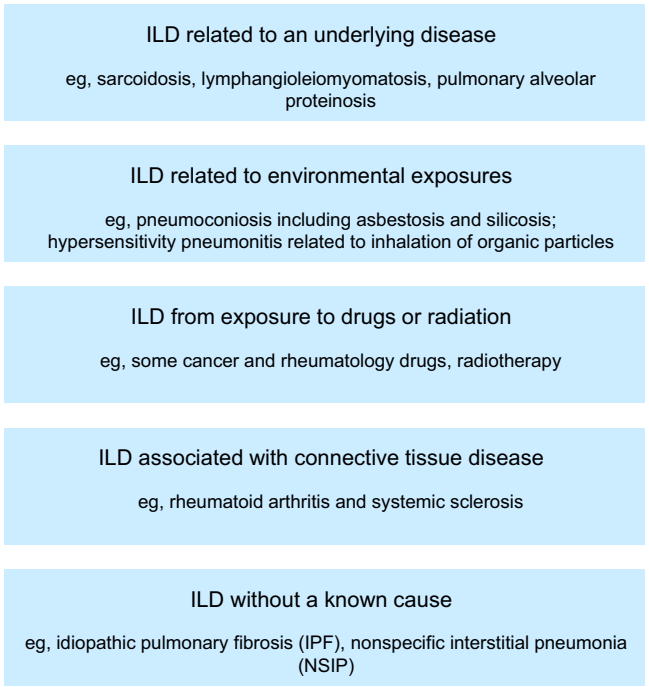

Figure 2. Clinical categories of interstitial lung diseases (ILDs). Adapted from Wijsenbeek et al.[2]

Comorbidities are common in people with ILD, including cardiovascular disease, lung cancer, obstructive sleep apnoea, gastro-oesophageal reflux, pulmonary hypertension and depression. Comorbidities adversely affect mortality and health-related quality of life in ILD, and thus it is important that these are identified and optimally treated. Musculoskeletal dysfunction is also prevalent in ILD. Quadriceps strength and endurance were found to be reduced by 20 to 30% in people with ILD compared with healthy controls[17] and strongly related to impaired exercise capacity. People with ILD took 65% fewer daily steps compared with their healthy age-matched peers,[18] and those with the lowest levels of physical activity had the worst health-related quality of life, independent of respiratory function.[19] There was a threefold increase in the hazard of death for patients with ILD who were highly sedentary ($<$ 3,300 steps/day).[19] Arthropathy is common in people with ILDs that are related to connective tissue disease (eg, rheumatoid arthritis, systemic sclerosis) and may have a profound impact on functional mobility and physical activity levels.

An acute exacerbation of IPF is defined as an acute, clinically significant respiratory deterioration with evidence of new widespread alveolar abnormality on high-resolution computed tomography. Acute exacerbations occur in 5 to 10% of patients with IPF annually.[20] An acute exacerbation of IPF is a life-threatening event, with in-hospital mortality of $>$ 50%, and median survival following an exacerbation of approximately 4 months.[20] It is now recognised that acute exacerbations also occur in non-IPF ILDs, although the prognosis may be more favourable. Amongst 102 patients with an acute exacerbation, 90-day mortality was 55% in those with IPF, compared with 31% in those with non-IPF ILDs.[21] Care needs in survivors are substantial, with 63% of those with IPF and 41% of those with non-IPF ILDs requiring long-term oxygen therapy at hospital discharge.

Physiotherapy management of ILD

The approval of anti-fibrotic drugs for people with IPF (pirfenidone and nintedanib) was a watershed moment, when IPF became a treatable disease for the first time.[22,23] The recent identification of the PF-ILD phenotype is also highly significant, as clinical trials have demonstrated that these patients have a similar response to anti-fibrotic treatments as is seen in IPF.[7] However, anti-fibrotic therapies only slow the progression of lung disease; they do not reverse the changes that have already occurred. In addition, there is no convincing evidence that these therapies improve symptoms or quality of life.[22,23] As a result, people with IPF and PF-ILD will likely live longer with a chronic respiratory disease that has a high burden of symptoms and disability. Physiotherapists make a critical contribution to the comprehensive management of people with ILD, with key treatment goals including reducing symptoms, improving exercise capacity, enhancing self-management and improving health-related quality of life.

Pulmonary rehabilitation

There is robust evidence that pulmonary rehabilitation delivers important benefits for people with ILD. A recent Cochrane review included 21 randomised controlled trials, of which 16 were included in a meta-analysis.[24] Outcomes of functional exercise capacity, dyspnoea and health-related quality of life are in Table 1. Improvement in 6-minute walk distance (6MWD) following pulmonary rehabilitation was clinically significant (mean difference to usual care 40 m), with the lower end of the confidence interval exceeding the minimum important difference. Results were similar in the subgroup of participants with IPF, providing reassurance that gains can be made

Table 1
Changes in functional exercise capacity, dyspnoea and health-related quality of life following pulmonary rehabilitation. From Dowman et al.[24]

Clinical measures	All interstitial lung disease		Idiopathic pulmonary fibrosis only	
	n	mean (95% CI)	n	mean (95% CI)
6-minute walk distance immediately after PR *(m)*	585	40 (33 to 47)	278	37 (26 to 48)
6-minute walk distance at 6 to 12-month follow-up *(m)*	297	32 (16 to 49)	123	2 (–25 to 28)
Dyspnoea immediately after PR (MMRC scale, SMD)	348	–0.36 (–0.58 to –0.14)	155	–0.41 (–0.74 to –0.09)
Dyspnoea at 6 to 12-month follow-up (MMRC scale, SMD)	335	–0.29 (–0.49 to –0.10)	123	–0.38 (–0.72 to –0.05)
SGRQ total score immediately after PR	478	–9 (–11 to –8)	194	–8 (–11 to –5)
SGRQ total score at 6 to 12-month follow-up	240	–5 (–8 to –2)	89	–3 (–7 to 1)

Data are change in outcome in randomised controlled trials of pulmonary rehabilitation compared to usual care.
ILD = interstitial lung disease, IPF = idiopathic pulmonary fibrosis, MMRC = modified Medical Research Council, PR = pulmonary rehabilitation, SGRQ = St George's Respiratory Questionnaire, SMD = standardised mean difference.

in this group of patients who often have progressive disease. Important improvements in dyspnoea and health-related quality of life were also evident, both for all participants with ILD and in the IPF subgroup. Importantly, the updated Cochrane review reported persistent benefits of pulmonary rehabilitation for the first time. Those who undertook pulmonary rehabilitation had a higher 6MWD, lower dyspnoea and better health-related quality of life 6 to 12 months following program completion. Within-group changes showed that these benefits were not necessarily 'maintained' at the same level in rehabilitation participants, but the control group had a more substantial deterioration over time, resulting in a sustained between-group difference.

Only four randomised trials have examined the impact of pulmonary rehabilitation on survival, with follow-up periods ranging from 6 to 12 months. Whilst the results tended to favour pulmonary rehabilitation, this estimate came with substantial uncertainty (OR for mortality 0.40, 95% CI 0.14 to 1.12, 291 participants).[24] A retrospective cohort study including 701 participants from five countries reported that a larger improvement in 6MWD following pulmonary rehabilitation was associated with a lower hazard ratio for death or lung transplant for both inpatient pulmonary rehabilitation (hazard ratio (HR) per 10 m = 0.94, 95% CI 0.91 to 0.97) and outpatient pulmonary rehabilitation (HR 0.97, 95% CI 0.95 to 1.00).[25] Participation in $\geq$ 80% of sessions was associated with a 33% lower risk of death (HR 0.67, 95% CI 0.49 to 0.92). Causality cannot be established by this uncontrolled study.

There is no evidence that those with more severe IPF cannot benefit from pulmonary rehabilitation, but it is apparent that more sustained benefits are observed earlier in the disease. Pulmonary function at enrolment (forced vital capacity, diffusion capacity for carbon monoxide) was found to not predict response to pulmonary rehabilitation.[26] Those with lower baseline 6MWD and worse dyspnoea had the greatest improvement at the end of the program, consistent with the critical role of pulmonary rehabilitation in improving functional capacity and reducing symptoms. However, those with better physiology were more likely to have sustained benefits 6 months after program completion, both for pulmonary function (each 100-ml increase in forced vital capacity was associated with 2 m greater 6MWD) and pulmonary hypertension (each 10-mmHg decrease in pulmonary artery systolic pressure was associated with 15 m greater 6MWD).[26] These results suggest that referral to pulmonary rehabilitation early in the disease course should be encouraged.

Components of pulmonary rehabilitation programs for people with ILD have been found to be similar to those for people with other lung diseases, including COPD.[24] Inpatient and outpatient pulmonary rehabilitation programs have been found to be effective,[25] with the model often determined by healthcare system organisation. Program length varied but was most commonly 8 to 12 weeks of outpatient sessions (two to three sessions per week). Exercise components included aerobic and resistance training. Unsurprisingly, greater benefits were achieved by participants in whom consistent progression of exercise intensity and duration occurred over the course of the program.[26] This is easier to achieve in those who attend a larger number of sessions and those who have less progressive disease.[27]

Delivery of an effective dose of exercise training can be particularly challenging in those who experience profound exercise-induced desaturation. Interval training may be a useful strategy with which to attenuate exertional desaturation in ILD. In nine people with ILD (mean forced vital capacity 66% predicted), oxyhaemoglobin saturation (SpO_2) was higher after 10 minutes of high-intensity interval training (30-second intervals, 100% peak work and passive rest) compared with moderate-intensity interval training (2-minute intervals, 80% peak work and 40% peak work) and moderate-intensity continuous training (60% peak work); however, exertional desaturation was not abolished (mean 87% versus 85% and 84%, respectively).[28] Oxygen supplementation during training is also critical for many patients with ILD, but standard methods may be insufficient to correct SpO_2 during exercise. Recent studies have shown that high-flow nasal cannula oxygen therapy increases exercise time, improves SpO_2 and reduces leg fatigue during exercise in IPF.[29] It is not yet known whether these strategies improve pulmonary rehabilitation outcomes.

Non-exercise components of pulmonary rehabilitation have included education, nutritional advice, stress management, breathing exercises, occupational therapy and social support.[24] Key educational topics for inclusion in pulmonary rehabilitation for ILD have been identified by patients and health professionals, using a consensus approach (Box 1).[30] Some of these topics require clinicians to have ILD-specific knowledge and expertise (eg, managing medication side-effects, managing cough). People with ILD were found to be generally comfortable attending pulmonary rehabilitation programs that included other people with a variety of lung conditions, but expected ILD-specific education to be provided.[31]

Despite the known benefits of pulmonary rehabilitation, many patients do not have the opportunity to undertake a program. Referral rates have been reported as ranging from 20 to 40% of eligible patients[32,33] which, although higher than rates commonly reported in other lung diseases, is far from optimal, considering that this treatment is recommended in clinical practice guidelines across the world. People with ILD face similar barriers to attending pulmonary rehabilitation, as reported in COPD, including fear of exercise, debilitating symptoms and the burden of travel to the pulmonary rehabilitation centre.[33] Delivery of pulmonary rehabilitation directly into the home using technology appears to be safe and may be beneficial to improve pulmonary rehabilitation access and outcomes for people with ILD, although few patients with ILD have been included in randomised trials to date.[34]

Musculoskeletal care

People with ILD related to underlying connective tissue disease frequently have arthropathy, which may impact physical function. Lung involvement is the most common extra-articular manifestation of rheumatoid arthritis, occurring in up to 60% of patients across the disease course.[35] As a result, a subgroup of patients with ILD who present for rehabilitation may have limitations to whole-body exercise related to joint disease, with associated reductions in range of motion, pain and disability. It is critical that optimal musculoskeletal

Box 1. Education topics for pulmonary rehabilitation in interstitial lung disease. From Holland et al.[30]

Core education topics
- Staying well with ILD: managing flare ups, regular vaccinations, importance of exercise, nutrition
- Keeping fit and strong after pulmonary rehabilitation
- Using oxygen therapy
- Managing breathlessness and cough
- Managing fatigue
- Managing anxiety, depression and panic

Optional education topics[a]
- End-of-life care and advance directives
- Accessing support for patients and carers
- Managing medications and side effects
- Tuning up the whole system – managing co-existing medical conditions

[a] To be delivered if local resources allow.

management is delivered, to facilitate participation in rehabilitation and regular physical activity. This may include specific muscle strengthening exercises, exercise to address functional limitations, modifications to aerobic training to reduce joint loading, and education regarding self-management of fatigue, pain and stress. Close collaboration between physiotherapists with expertise in rheumatology, orthopaedics and respiratory management may be required.

Supplemental oxygen therapy

Hypoxaemia occurring during rest, sleep or exercise is a cardinal feature of the ILDs. With the exception of lung transplantation, supplemental oxygen is the only treatment that improves hypoxaemia that persists despite optimal medical management.[36] The principles underlying delivery of supplemental oxygen are similar in ILD and other chronic lung diseases; however, the greater magnitude of hypoxaemia (and hence greater oxygen requirements) experienced by people with ILD often poses unique challenges for delivery.

The American Thoracic Society (ATS) clinical practice guideline on home oxygen therapy for adults with chronic lung disease[36] provides a strong recommendation for long-term oxygen therapy for at least 15 hours/day in people with ILD who have severe chronic resting room air hypoxaemia ($PaO_2 \leq 55$ mmHg). The recommendation is based on indirect evidence from clinical trials in COPD demonstrating a mortality benefit, and ethical concerns about withholding oxygen from patients with ILD who may be profoundly hypoxaemic and dyspnoeic at rest. Long-term oxygen therapy is a well-accepted treatment for people with advanced ILD and is commonly available in jurisdictions with supplemental oxygen programs. There are few data on the clinical implications of hypoxaemia occurring only during sleep, or the impact of its treatment; as a result, isolated sleep-induced hypoxaemia is not generally considered to be an indication for oxygen therapy in ILD.

For patients who experience isolated exertional hypoxemia ($SpO_2 \leq 88\%$) the ATS guideline makes a conditional recommendation for treatment with ambulatory oxygen, based on low-quality evidence.[36] Most studies that underpin the recommendation evaluated the acute effects of oxygen on exercise performance during a single session in the laboratory, showing consistent improvements in 6MWD and exercise endurance, and reduced dyspnoea during exercise testing. However, it is unclear whether these acute effects of ambulatory oxygen are translated into beneficial effects during daily life. A crossover randomised trial with a 2-week treatment period compared ambulatory oxygen with no treatment in participants with fibrotic ILD and showed a significant improvement in the primary outcome of health-related quality of life.[37] However, participants and assessors were not blinded to treatment allocation, and longer-term effects could not be evaluated. The conditional recommendation for this treatment acknowledges the potential for negative impacts of ambulatory oxygen therapy, including cumbersome and complicated equipment, perceived stigma, unmet expectations for symptom relief (particularly dyspnoea), reduced independence, and increased caregiver burden.[36] As a result of these potential burdens some patients may choose not to use ambulatory oxygen therapy, and shared decision-making between patients and healthcare professionals is required. The availability of ambulatory oxygen and the criteria for its prescription vary widely across jurisdictions, partly reflecting the limited evidence of benefit underpinning its use.

Long-term oxygen therapy is typically delivered using a stationary oxygen concentrator in the home. Ambulatory oxygen therapy can be delivered using a variety of portable devices with different characteristics and costs. Metal oxygen cylinders are available in multiple sizes and are relatively low in cost. A typical oxygen tank that is pulled on a trolley (E tank in the USA) lasts less than 2 hours if high flow rates (6 l/minute) are required, which will limit ambulation outside the home. Oxygen conserving devices may be used to prolong the duration of supply, but often do not deliver a sufficient oxygen dose for those with higher requirements such as in ILD. Multiple tanks may be required for patients requiring flow rates > 3 l/minute in order to spend more than 2 to 4 hours away from home. Portable concentrators are battery powered (and thus do not require refilling) and generally lighter than metal cylinders. However, portable concentrators are expensive (up to US$4,000); the pulsed dose of oxygen delivered may be insufficient for patients with higher oxygen requirements; and the battery runs out more quickly at higher settings and respiratory rates. Neither delivery system fully corrects oxyhaemoglobin saturation on exertion in ILD, even on maximal settings.[38] Portable liquid oxygen allows delivery of continuous flow oxygen up to 15 l/minute, and enables a longer duration of use. However portable liquid oxygen is not available in many jurisdictions due to cost (up to four times higher per patient compared with cylinders or portable concentrators). The increased cost for liquid oxygen is primarily attributable to the need for specialised delivery vehicles and frequent refilling of home reservoirs.

Optimisation of oxygen therapy is a key role for physiotherapists in ILD care. Patients should be prescribed the ambulatory oxygen delivery device that best meets their physical, physiological and lifestyle needs, and this should be re-evaluated as disease progresses. For instance, some patients who have lower oxygen requirements or are very active may be best served by small, lightweight cylinders or a portable concentrator that can be carried in a backpack. Disease progression and increased oxygen requirements may require patients to transition from portable concentrators to metal cylinders, or even multiple cylinders. Consideration of how the ambulatory oxygen device can best be transported by patients and caregivers is critical, including use of trolleys and wheeled walkers. Multiple stationary concentrators are sometimes needed to meet the oxygen requirements of patients on long-term oxygen therapy with very advanced disease.

Patients frequently report a lack of information about how to use their oxygen equipment and have low confidence in their skills.[39] All patients and caregivers should receive instruction and training in the safe and effective use of their oxygen equipment.[36] Important considerations for safety, education, training and monitoring in patients prescribed oxygen therapy are presented in Table 2.

Education, self-management and support

Supportive measures, including education and psychological support, are key components of comprehensive ILD care (Figure 1)[1] and are a high priority for patients. In a systematic review of supportive care needs in pulmonary fibrosis,[40] including data from 2,621 participants and 590 caregivers, the need for more information and education was reported in 26 of 35 studies. Specific information needs reported by participants included understanding disease progression and prognosis; oxygen therapy, including travel with oxygen; managing side-effects of drug therapies; planning for end-of-life care; coping strategies; and managing symptoms of breathlessness

Table 2
Considerations for safety, education, training and monitoring in patients prescribed oxygen therapy. Adapted from Jacobs et al.[36]

Category	Considerations
Safety	• Education regarding avoidance of trips and falls; decreasing fire risk by not smoking or allowing smoking in the home; avoidance of open flames or sparks; use of nonpetroleum nasal products • Instruct liquid-oxygen users on avoidance of skin burns from contact with frosted parts on liquid-oxygen-device connectors • Provide guidance on transporting and travelling safely with oxygen • Confirm the presence of back-up devices for emergencies or power loss
Smoking	• Instruct current smokers or caregiver smokers on smoking cessation and treatment of tobacco dependence; refer to appropriate resources • Alert patients and caregivers that use of e-cigarettes, or vaping, is associated with burn accidents in people receiving home oxygen therapy
Education and training	• Tailor patients' education to their health literacy and cultural contexts • Incorporate effective evaluation and return demonstration of their ability to use their prescribed devices both in the home and in ambulatory settings • Instruct patients and caregivers on troubleshooting equipment problems • Consider access to appropriate equipment on the basis of patients' physical, physiologic, and lifestyle/mobility needs
Monitoring	• Reassess patients' oxygen needs, with a frequency varying according to disease characteristics, such as rate of progression. • Reassess oxygen needs for patients who are newly prescribed oxygen after hospital discharge, to confirm ongoing oxygen requirements • Advise patients to bring their portable device to healthcare visits to assess its effectiveness and to reinforce self-management

and cough. Physiotherapists may address some of these informational needs in pulmonary rehabilitation programs, but there may be other opportunities including outpatient clinics, oxygen therapy clinics, support groups, patient seminars, and in the inpatient setting.

Self-management is a relatively new concept in ILD care, driven by changes to patient and healthcare professional expectations of treatments and outcomes in the anti-fibrotic era. Whereas previously it was assumed that patients with PF-ILD and IPF would face inexorable disease progression and death, with few treatment options or opportunities to be actively involved in their care, this is no longer the case. The focus of care has shifted to supporting individuals to live well with ILD, including strategies that can be employed by patients to maintain their health and wellbeing over time. Key elements of self-management for ILD identified by patients and healthcare professionals include exercise, physical activity, nutrition, weight management, regular vaccinations, avoiding infections, recognising deterioration, seeking help, managing symptoms, managing treatments, managing treatment side effects, and maintaining mental health.[41] The impact of self-management on clinical outcomes in ILD is not yet known.

In addition to the support provided by healthcare professionals, patients with ILD value opportunities for peer support. The ILDs are relatively rare conditions, so many individuals will not have met another person with the same diagnosis. Peer support programs are most commonly offered by patient organisations, often online or via telephone, and provide unique opportunities to share experiences and offer mutual support.[42] Linking individuals with ILD with patient organisations also provides a mechanism by which they can access patient-centred, up-to-date information to improve disease knowledge and self-management.

Symptom management and palliative care

Symptom management and palliative care aim to improve health-related quality of life and support individuals to live well with ILD. These interventions should not be restricted to end-of-life care, but are relevant across the disease course to relieve suffering and enhance wellbeing.[43] Whilst the evidence underpinning symptom management approaches is limited, a small number of randomised trials suggest that there may be important benefits for people with ILD. In 105 people with refractory breathlessness, 19 of whom had ILD, a multidisciplinary breathlessness support service significantly improved survival at 6 months compared with usual care.[44] In 53 patients with advanced fibrotic ILD, a community case conference to address palliative care needs improved symptoms and health-related quality of life after 4 weeks compared with usual care.[45] An intervention designed to reduce symptom burden and improve health-related quality of life in patient-caregiver dyads (n = 76) improved knowledge, confidence in managing disease and completion of advance care plans.[46] These interventions are individualised according to the needs and goals of patients, and may include both pharmacological (eg, low-dose opioids) and non-pharmacological components. Non-pharmacological intervention components included in recent clinical trials are in Box 2, many of which are within the scope of practice of physiotherapists. Many of these components aim to reduce breathlessness, but strategies to manage fatigue, cough and anxiety are also important.

Cough is a particularly distressing symptom for many people with ILD. Whilst there is little research to guide physiotherapy management of cough, strategies that may be helpful include avoidance of cough triggers (eg, dusty environments, animal fur, cleaning products, perfumes); regularly sipping water and maintaining good hydration; using an air humidifier at home; use of non-medicated lozenges; nose breathing to warm and moisten inspired air; controlled breathing exercises, including during exercise if this is a trigger; mindfulness and distraction techniques; regular swallowing and avoidance of throat clearing; and airway clearance techniques if a productive cough is present.

Inpatient care

Physiotherapy management for inpatients with ILD may be required if the admission is for management of an acute exacerbation, although treatment options are limited. There are no proven therapies for acute exacerbations of ILD, with most centres delivering high-dose corticosteroids and antibiotics, as well as respiratory support (oxygen therapy, non-invasive or invasive mechanical ventilation).[47] Marked exercise limitation and increased long-term oxygen requirements are common in those who survive acute exacerbations, and these patients may benefit from physiotherapy management to optimise function and independence.

Transplant preparation and rehabilitation

Lung transplantation is a life-saving and life-prolonging procedure for people with end-stage fibrosing ILD. Recent years have seen an increase in the proportion of lung transplants performed for ILD,[48] with a concomitant increase in the number of recipients requiring long-term management. Physiotherapists play a key role in optimal management of patients with ILD before and after lung transplantation.

Physical rehabilitation is considered a core component of preparation for lung transplantation and often continues throughout the waiting period. Standard approaches to pulmonary rehabilitation can be applied, with modifications for disease severity, disease progression, functional deterioration and comorbidities.[49] There is some evidence that candidates who underwent rehabilitation had improvements in 6MWD and health-related quality of life, as well as reduced hospital length of stay and improved survival following transplantation in patients with IPF;[50] however, this is largely from uncontrolled studies. Rehabilitation also provides an opportunity for pre-transplant education, including preparation for the perioperative period (secretion management, controlled coughing, pain management, importance of early mobilisation) and optimisation of disease management (oxygen therapy, pacing, energy conservation, regular exercise).[51]

During the acute inpatient phase following transplantation, physiotherapy management (in intensive care and on the ward) comprises

Box 2. Components of symptom management and palliative care interventions for interstitial lung disease. Components are from randomised controlled trials of symptom management and palliative care interventions.[44–46]

- Relaxation and breathing control
- Cough control strategies
- Handheld fan
- Oxygen therapy
- Crisis plan for breathlessness
- Energy conservation and pacing
- Walking aids, home adaptations
- Exercise training, pulmonary rehabilitation
- Advance care planning
- Education and information
- Counselling
- Cognitive behavioural therapy
- Education and support for caregivers
- Multidisciplinary case conference and care planning
- Multidisciplinary breathlessness support service

respiratory management, including airway clearance and oxygen titration, early mobility, exercise training and education.[52] Rehabilitation in the early post-transplant phase (months 1 to 6) may occur in the inpatient or outpatient setting and includes aerobic, resistance and flexibility training.[49] Clinical trials have demonstrated benefits of this approach, including improved 6MWD and quadriceps strength, increased daily physical activity, and improved bone mineral density. Rehabilitation may also be important in long-term management (> 6 months), to improve exercise capacity, strength and physical activity levels, and to aid in the management of transplant-related comorbidities such as hypertension, hyperlipidaemia and diabetes.[49]

Future directions for research and practice

It is an exciting time to be involved in ILD research and clinical practice, with advances in disease-modifying treatments for IPF and PF-ILD that bring hope for improved patient outcomes. In this context, physiotherapy interventions to maximise long-term health and wellbeing are increasingly important and expanding in scope.

Whilst pulmonary rehabilitation is a core component of ILD care that generally delivers excellent patient outcomes, it is clear that some patients do not benefit as expected.[27] Future research should examine whether alternative training strategies (including high-intensity interval training and high-flow oxygen therapy) can optimise short-term and longer-term outcomes, and in whom such strategies should be applied. It will be important to quantify the benefits of repeating pulmonary rehabilitation, including health economic outcomes, to inform future decisions on funding and policy. The emerging field of self-management in ILD provides significant research opportunities to define its components, and test outcomes in clinical trials. Similarly, models of symptom management such as breathlessness clinics show great potential, but require further rigorous testing and a clearer understanding of the essential components. There are currently no evidence-based strategies for managing cough in ILD, and there is a pressing need for research in this area. There is enormous scope for research to address knowledge gaps regarding oxygen therapy for ILD, including an urgent need for development and testing of improved oxygen delivery systems that can better meet the needs of this patient group who may have profound hypoxaemia. As ILD is relatively uncommon, acceleration of clinical research across all fields will require innovative designs that enable participation by patients who are located away from major centres (eg, teletrials) and master protocols that allow inclusion of well-phenotyped patients with a variety of ILD subtypes. Physiotherapists can support research in ILD by encouraging patients to participate in national ILD registries and clinical trials, both of which will be required to advance knowledge and expand treatment options for these debilitating conditions.

In clinical practice, there are many opportunities to improve access to the best care for people with ILD. Whilst it is well accepted that a multidisciplinary team is needed for accurate ILD diagnosis[15] and efforts are well underway to ensure that patients have access to expert diagnostic services regardless of geography,[53] access to the full range of services required for ILD care remains challenging, particularly for those in regional areas.[54] Optimal models of specialist ILD care in the future should include access to multidisciplinary team management, with capacity to deliver care via telehealth to ensure that this expertise is available to all patients. Recent developments in telerehabilitation may facilitate access to this critical component of ILD care, and similar models could be explored for delivery of self-management, symptom management and palliative care interventions. Finally, there is a need to increase awareness amongst physiotherapists and other healthcare professionals regarding the burden of ILD, the availability of effective treatments and opportunities to optimise outcomes for this patient group.

eAddenda: Nil.

Ethics approval: Not applicable.

Competing interests: Nil.

Source(s) of support: Nil.

Acknowledgements: Nil.

Provenance: Invited. Peer reviewed.

Correspondence: Anne E Holland, Departments of Physiotherapy and Respiratory Medicine, Alfred Health, Melbourne, Australia. Email: a.holland@alfred.org.au

References

1. Wijsenbeek MS, Holland AE, Swigris JJ, Renzoni EA. Comprehensive Supportive Care for Patients with Fibrosing Interstitial Lung Disease. *Am J Respir Crit Care Med*. 2019;200:152–159.
2. Wijsenbeek M, Cottin V. Spectrum of Fibrotic Lung Diseases. *N Engl J Med*. 2020;383:958–968.
3. McGroder CF, Zhang D, Choudhury MA, Salvatore MM, D'Souza BM, Hoffman EA, et al. Pulmonary fibrosis 4 months after COVID-19 is associated with severity of illness and blood leucocyte telomere length. *Thorax*. 2021;76:1242–1245.
4. Zhang P, Li J, Liu H, Han N, Ju J, Kou Y, et al. Long-term bone and lung consequences associated with hospital-acquired severe acute respiratory syndrome: a 15-year follow-up from a prospective cohort study. *Bone Res*. 2020;8:8.
5. Global Burden of Disease Chronic Respiratory Disease Collaborators. Prevalence and attributable health burden of chronic respiratory diseases, 1990–2017: a systematic analysis for the Global Burden of Disease Study 2017. *Lancet Resp Med*. 2020;8:585–596.
6. Zheng Q, Cox IA, Campbell JA, Xia Q, Otahal P, de Graaff B, et al. Mortality and survival in idiopathic pulmonary fibrosis: a systematic review and meta-analysis. *ERJ Open Res*. 2022;8.
7. Cottin V, Teague R, Nicholson L, Langham S, Baldwin M. The Burden of Progressive-Fibrosing Interstitial Lung Diseases. *Front Med (Lausanne)*. 2022;9:799912.
8. King TE, Jr., Lee JS. Cryptogenic Organizing Pneumonia. *N Engl J Med*. 2022;386:1058–1069.
9. Carvajalino S, Reigada C, Johnson MJ, Dzingina M, Bajwah S. Symptom prevalence of patients with fibrotic interstitial lung disease: a systematic literature review. *BMC Pulm Med*. 2018;18:78.
10. Swigris JJ, Stewart AL, Gould MK, Wilson SR. Patients' perspectives on how idiopathic pulmonary fibrosis affects the quality of their lives. *Health Qual Life Outcomes*. 2005;3:61.
11. Yount SE, Beaumont JL, Chen SY, Kaiser K, Wortman K, Van Brunt DL, et al. Health-Related Quality of Life in Patients with Idiopathic Pulmonary Fibrosis. *Lung*. 2016;194:227–234.
12. Klein S, Logan A, Lindell KO. A scoping review of unmet needs of caregivers of patients with pulmonary fibrosis. *Curr Opin Support Palliat Care*. 2021;15:226–232.
13. Wong AW, Koo J, Ryerson CJ, Sadatsafavi M, Chen W. A systematic review on the economic burden of interstitial lung disease and the cost-effectiveness of current therapies. *BMC Pulm Med*. 2022;22:148.
14. Collard HR, Chen SY, Yeh WS, Li Q, Lee YC, Wang A, et al. Health care utilization and costs of idiopathic pulmonary fibrosis in U.S. Medicare beneficiaries aged 65 years and older. *Ann Am Thorac Soc*. 2015;12:981–987.
15. Prasad JD, Mahar A, Bleasel J, Ellis SJ, Chambers DC, Lake F, et al. The interstitial lung disease multidisciplinary meeting: A position statement from the Thoracic Society of Australia and New Zealand and the Lung Foundation Australia. *Respirology*. 2017;22:1459–1472.
16. Lama VN, Flaherty KR, Toews GB, Colby TV, Travis WD, Long Q, et al. Prognostic value of desaturation during a 6-minute walk test in idiopathic interstitial pneumonia. *Am J Respir Crit Care Med*. 2003;168:1084–1090.
17. Mendoza L, Gogali A, Shrikrishna D, Cavada G, Kemp SV, Natanek SA, et al. Quadriceps strength and endurance in fibrotic idiopathic interstitial pneumonia. *Respirology*. 2014;19:138–143.

18. Wallaert B, Monge E, Le Rouzic O, Wemeau-Stervinou L, Salleron J, Grosbois JM. Physical activity in daily life of patients with fibrotic idiopathic interstitial pneumonia. *Chest*. 2013;144:1652–1658.
19. Bahmer T, Kirsten AM, Waschki B, Rabe KF, Magnussen H, Kirsten D, et al. Clinical Correlates of Reduced Physical Activity in Idiopathic Pulmonary Fibrosis. *Respiration*. 2016;91:497–502.
20. Raghu G, Collard HR, Egan JJ, Martinez FJ, Behr J, Brown KK, et al. An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management. *Am J Respir Crit Care Med*. 2011;183:788–824.
21. Miyashita K, Kono M, Saito G, Koyanagi Y, Tsutsumi A, Kobayashi T, et al. Prognosis after acute exacerbation in patients with interstitial lung disease other than idiopathic pulmonary fibrosis. *Clin Respir J*. 2021;15:336–344.
22. King TE, Jr., Bradford WZ, Castro-Bernardini S, Fagan EA, Glaspole I, Glassberg MK, et al. A phase 3 trial of pirfenidone in patients with idiopathic pulmonary fibrosis. *N Engl J Med*. 2014;370:2083–2092.
23. Richeldi L, du Bois RM, Raghu G, Azuma A, Brown KK, Costabel U, et al. Efficacy and safety of nintedanib in idiopathic pulmonary fibrosis. *N Engl J Med*. 2014;370:2071–2082.
24. Dowman L, Hill CJ, May A, Holland AE. Pulmonary rehabilitation for interstitial lung disease. *Cochrane Database Syst Rev*. 2021;2:CD006322.
25. Guler SA, Hur SA, Stickland MK, Brun P, Bovet L, Holland AE, et al. Survival after inpatient or outpatient pulmonary rehabilitation in patients with fibrotic interstitial lung disease: a multicentre retrospective cohort study. Thorax. Published online ahead of print, August 30 2021; https://doi.org/10.1136/thoraxjnl-2021-217361.
26. Dowman LM, McDonald CF, Hill CJ, Lee AL, Barker K, Boote C, et al. The evidence of benefits of exercise training in interstitial lung disease: a randomised controlled trial. *Thorax*. 2017;72:610–619.
27. Nakazawa A, Dowman LM, Cox NS, McDonald CF, Hill CJ, Lee AL, et al. Factors predicting progression of exercise training loads in people with interstitial lung disease. *ERJ Open Res*. 2019;5.
28. Dowman LM, May AK, Cox NS, Morris NR, Nakazawa A, Parker L, et al. Attenuation of exertional desaturation and preference for interval exercise compared to continuous exercise in people with interstitial lung disease. *Respirology*. 2021;26:1076–1079.
29. Harada J, Nagata K, Morimoto T, Iwata K, Matsunashi A, Sato Y, et al. Effect of high-flow nasal cannula oxygen therapy on exercise tolerance in patients with idiopathic pulmonary fibrosis: A randomized crossover trial. *Respirology*. 2022;27:144–151.
30. Holland AE, Watson A, Glaspole I. Comprehensive pulmonary rehabilitation for interstitial lung disease: a consensus approach to identify core education topics. *Patient Educ Couns*. 2019;102:1135–1130.
31. Holland AE, Fiore JF, Jr., Goh N, Symons K, Dowman L, Westall G, et al. Be honest and help me prepare for the future: What people with interstitial lung disease want from education in pulmonary rehabilitation. *Chronic Respir Dis*. 2015;12:93–101.
32. de Andrade JA, Kulkarni T, Neely ML, Hellkamp AS, Case AH, Guntupalli K, et al. Implementation of guideline recommendations and outcomes in patients with idiopathic pulmonary fibrosis: Data from the IPF-PRO registry. *Respir Med*. 2021;189:106637.
33. Hoffman M, Mellerick C, Symons K, Glaspole I, Holland AE. Pulmonary rehabilitation for interstitial lung disease: Referral and patient experiences. *Chronic Respir Dis*. 2021;18:14799731211046022.
34. Cox NS, McDonald CF, Mahal A, Alison JA, Wootton R, Hill CJ, et al. Telerehabilitation for chronic respiratory disease: a randomised controlled equivalence trial. Thorax. Published online ahead of print, Oct 14 2021; https://doi.org/10.1136/thoraxjnl-2021-216934.
35. Kadura S, Raghu G. Rheumatoid arthritis-interstitial lung disease: manifestations and current concepts in pathogenesis and management. *Eur Respir Rev*. 2021;30.
36. Jacobs SS, Krishnan JA, Lederer DJ, Ghazipura M, Hossain T, Tan AM, et al. Home Oxygen Therapy for Adults with Chronic Lung Disease. An Official American Thoracic Society Clinical Practice Guideline. *Am J Respir Crit Care Med*. 2020;202:e121–e141.
37. Visca D, Montgomery A, de Lauretis A, Sestini P, Soteriou H, Maher TM, et al. Ambulatory oxygen in interstitial lung disease. *Eur Respir J*. 2011;38:987–990.
38. Khor YH, Hazard A, Symons K, Westall G, Glaspole I, Goh N, et al. Portable Oxygen Concentrators versus Oxygen Cylinder during Walking in Interstitial Lung Disease: A Randomized Crossover Trial. *Respirology*. 2016;22:1598–1603.
39. Khor YH, Goh NSL, McDonald CF, Holland AE. Oxygen Therapy for Interstitial Lung Disease: A Mismatch Between Patient Expectations and Experiences. *Ann Am Thorac Soc*. 2017;14:888–895.
40. Lee JYT, Tikellis G, Corte TJ, Goh NS, Keir GJ, Spencer L, et al. The supportive care needs of people living with pulmonary fibrosis and their caregivers: a systematic review. *Eur Respir Rev*. 2020;29.
41. Lee JYT, Tikellis G, Glaspole I, Khor YH, Symons K, Holland AE. Self-management for pulmonary fibrosis: Insights from people living with the disease and healthcare professionals. *Patient Educ Couns*. 2022;105:956–964.
42. Tikellis G, Lee JYT, Corte TJ, Maloney J, Bartlett M, Crawford T, et al. Peer Connect Service for people with pulmonary fibrosis in Australia: Participants' experiences and process evaluation. *Respirology*. 2020;25:1053–1059.
43. Kreuter M, Bendstrup E, Russell AM, Bajwah S, Lindell K, Adir Y, et al. Palliative care in interstitial lung disease: living well. *Lancet Respir Med*. 2017;5:968–980.
44. Higginson IJ, Bausewein C, Reilly CC, Gao W, Gysels M, Dzingina M, et al. An integrated palliative and respiratory care service for patients with advanced disease and refractory breathlessness: a randomised controlled trial. *Lancet Respir Med*. 2014;2:979–987.
45. Bajwah S, Ross JR, Wells AU, Mohammed K, Oyebode C, Birring SS, et al. Palliative care for patients with advanced fibrotic lung disease: a randomised controlled phase II and feasibility trial of a community case conference intervention. *Thorax*. 2015;70:830–839.
46. Lindell KO, Klein SJ, Veatch MS, Gibson KF, Kass DJ, Nouraie M, et al. Nurse-Led Palliative Care Clinical Trial Improves Knowledge and Preparedness in Caregivers of Patients with Idiopathic Pulmonary Fibrosis. *Ann Am Thorac Soc*. 2021;18:1811–1821.
47. Polke M, Kondoh Y, Wijsenbeek M, Cottin V, Walsh SLF, Collard HR, et al. Management of Acute Exacerbation of Idiopathic Pulmonary Fibrosis in Specialised and Non-specialised ILD Centres Around the World. *Front Med (Lausanne)*. 2021;8:699644.
48. Chambers DC, Perch M, Zuckermann A, Cherikh WS, Harhay MO, Hayes D, Jr., et al. The International Thoracic Organ Transplant Registry of the International Society for Heart and Lung Transplantation: Thirty-eighth adult lung transplantation report - 2021; Focus on recipient characteristics. *J Heart Lung Transplant*. 2021;40:1060–1072.
49. Wickerson L, Rozenberg D, Janaudis-Ferreira T, Deliva R, Lo V, Beauchamp G, et al. Physical rehabilitation for lung transplant candidates and recipients: An evidence-informed clinical approach. *World J Transplant*. 2016;6:517–531.
50. Florian J, Rubin A, Mattiello R, Fontoura FF, Camargo Jde J, Teixeira PJ. Impact of pulmonary rehabilitation on quality of life and functional capacity in patients on waiting lists for lung transplantation. *J Bras Pneumol*. 2013;39:349–356.
51. Rochester CL, Fairburn C, Crouch RH. Pulmonary rehabilitation for disorders other than chronic obstructive pulmonary disease. *Clin Chest Med*. 2014;35:369–389.
52. Tarrant BJ, Holland A, Le Maitre C, Robinson R, Corbett M, Bondarenko J, et al. The timing and extent of acute physiotherapy involvement following lung transplantation: An observational study. *Physiother Res Int*. 2018;23:e1710.
53. Mackintosh JA, Glenn L, Barnes H, Dunn E, Bancroft S, Reddy T, et al. Benefits of a virtual interstitial lung disease multidisciplinary meeting in the face of COVID-19. *Respirology*. 2021;26:612–615.
54. Tikellis G, Corte TJ, Teoh AKY, Glaspole IN, Macansh S, Holland AE. Barriers and facilitators to best care for idiopathic pulmonary fibrosis in Australia. *Respirology*. 2022;27:76–84.