The term interstitial lung diseases (ILDs) encompasses a large and heterogeneous group of disorders. Some ILDs have a known cause, including ILDs associated with autoimmune rheumatic diseases (also known as connective tissue diseases) such as rheumatoid arthritis, systemic sclerosis (SSc), myositis, or Sjögren syndrome; with sarcoidosis; with exposure to organic antigens such as mold or feathers (hypersensitivity pneumonitis); or with exposure to toxic material such as asbestos. ILDs may also develop without an identified cause or be unclassifiable. The most well-known ILD is idiopathic pulmonary fibrosis (IPF), which is, by definition, a progressive fibrosing disease of unknown cause. Other ILDs may also develop a progressive fibrosing phenotype characterized by increasing fibrotic abnormalities on scans of the lungs, decline in lung function, worsening symptoms and quality of life, and early mortality (see Types of ILDs that may be associated with a progressive fibrosing phenotype). In this article, we discuss the diagnosis and management of fibrosing ILDs and how NPs can help support patients with these diseases.

## Diagnosis and monitoring of ILDs

ILDs with similar clinical and radiologic presentations may differ in etiology and clinical course and require different management strategies. It is critical to develop an accurate differential diagnosis for the patient’s ILD to ensure that the patient receives appropriate care and support. A diagnosis of ILD should be made based on the patient’s medical history, symptoms,
Management and support of patients with fibrosing interstitial lung diseases

pulmonary function tests, serologies, thorough assessment of environmental exposures, and expert review of a high-resolution computed tomography (HRCT) scan of the lungs.\(^4\) If the clinical and radiographic data are not sufficient to identify the subtype of ILD, a surgical lung biopsy may be performed to improve diagnostic clarity. In making a diagnosis of hypersensitivity pneumonitis, bronchoalveolar lavage may also be useful.\(^7\) Patients should be referred for an HRCT scan if they are suspected to have ILD. This includes patients over 60 years of age with unexplained chronic dyspnea or cough (who may have IPF), younger patients with autoimmune diseases who have respiratory symptoms, individuals with a family history of ILD, and patients receiving medication with the potential for pulmonary toxicity.\(^4\) ILD is a common manifestation of SSc and experts recommend that all patients diagnosed with SSc be screened for ILD using HRCT.\(^6,7\) It is also important to keep ILD on the differential for patients with pulmonary symptoms who do not respond to standard treatment for chronic obstructive pulmonary disease and asthma. All patients suspected to have ILD based on symptoms, inspiratory crackles on exam, or abnormal pulmonary function tests or imaging should be referred to a pulmonary specialist for evaluation.

The gold standard is that a diagnosis of ILD be made following multidisciplinary discussion (MDD) of the available data, although in clear-cut cases, a pulmonologist may not require an MDD to make a diagnosis of IPF. An MDD may be held virtually if it is challenging to convene a face-to-face meeting and should include a pulmonologist, a radiologist, and, where relevant, a rheumatologist and/or a pathologist.\(^4\) In a recent review of MDD records from 300 patients with ILDs in Canada, MDD led to a change in diagnosis in 40% of patients.\(^8\) The involvement of a radiologist is important not only to ensure that the most accurate diagnosis is made, but also to assess the fibrotic pattern seen on HRCT, as patients with a usual interstitial pneumonia pattern on HRCT have a faster rate of decline in lung function and higher mortality than patients with other patterns on HRCT.\(^9,10\) In circumstances where a patient does not fulfill the criteria for any specific ILD diagnosis even after MDD, a “working diagnosis” may be made to inform management; this should be revisited at regular intervals.\(^4\) These patients may also be considered to have unclassifiable ILD.

Fibrosing ILDs have a variable clinical course. Regular monitoring of patients with ILDs is important to assess disease progression and inform patient care and counseling. IPF is always progressive, but the rate at which it progresses is variable and patients may suffer acute deteriorations in lung function, referred to as acute exacerbations, which usually require hospitalization and are associated with very high mortality.\(^11,12\) It is important to note that patients with IPF who have preserved lung function at diagnosis are not protected from decline in lung function, or acute exacerbations, over the following year.\(^13,14\) Other fibrosing ILDs are not always progressive, but may develop a progressive phenotype associated with poor outcomes.\(^7\) While there is no standard definition for progression of ILD, an increase in fibrotic abnormalities on HRCT and/or a decline in forced vital capacity (FVC) of 10% or more of the predicted value are associated with a higher risk of mortality.\(^15-17\) Recent data from the INBUILD trial, which enrolled patients with fibrosing ILDs other than IPF who had demonstrated progression of ILD over the previous 2 years, showed that the rate of decline in FVC over the next year was the same in these patients as in patients with IPF.\(^18\) Monitoring patients with ILDs should include

Regular monitoring of patients with ILDs is important to assess disease progression and inform patient care and counseling.
pulmonary function tests, assessment of symptoms, and, where appropriate, repeat HRCT scans. The 6-minute walk test can be useful to monitor changes in exercise capacity and exertional oxygen desaturation, but its results should be interpreted with care, particularly in patients who have other manifestations of autoimmune disease or comorbidities that may affect exertional capacity, such as pulmonary hypertension, or pulse oximeter accuracy (for example Raynaud’s phenomenon).

The unpredictable course of ILDs makes it challenging for clinicians to explain the implications of an ILD diagnosis to patients. Nonetheless, it is important that NPs and other clinicians provide patients with information about the clinical course typically associated with their specific diagnosis and address their questions about the severity, progression, and possible future course of their ILD as clearly as possible. The specific diagnosis that a patient has, and its implications for prognosis and management, will likely require reiteration over several visits. It is important that clinicians utilize active listening and effective communication skills when providing information that may be misunderstood or distressing for patients. NPs have a key role to play in helping patients understand the tests that they undergo during disease diagnosis and monitoring, treatment options, best ways to manage the disease, and the support that is available to them (see Key points to communicate to patients with fibrosing ILDs).

Management of ILDs
Management of ILDs is dependent upon the diagnosis (or working diagnosis). For patients with hypersensitivity pneumonitis, avoiding exposure to the inciting antigen is fundamental to improving outcomes. IPF is a relentlessly progressive disease with a poor prognosis, and patients should receive prompt treatment with an approved antifibrotic therapy to slow disease progression. For patients with other fibrosing ILDs, the initiation and escalation of pharmacotherapy should be considered on an individual basis, based on disease severity, risk factors for and evidence of...
progression, other manifestations of the disease, and
the preferences of the patient. When the best course
of treatment is in question, management decisions
should be made based on MDD. Immunomodulatory
therapies may be prescribed by rheumatologists or
pulmonologists, while it is generally pulmonologists
who prescribe antifibrotic therapies.

**Immunosuppressants.** Immunosuppression is the
cornerstone of treatment for autoimmune diseases, as
it addresses the immune dysfunction characteristic of
these diseases. However, evidence for the effectiveness
of immunosuppression in slowing the progression of
ILD is limited. The results from randomized controlled
trials support the use of mycophenolate mofetil and
cyclophosphamide in the treatment of SSC-associated
ILD (SSc-ILD)\(^ {26-28}\). In Scleroderma Lung Study (SLS)
I, patients treated with oral cyclophosphamide had
a smaller mean decline in FVC than patients in the
placebo group after 1 year.\(^ {27}\) In SLS II, changes in FVC
were similar between patients treated with mycophe-
nolate mofetil for 2 years and those treated with oral
cyclophosphamide for 1 year followed by placebo for
1 year.\(^ {28}\) In a recent Delphi consensus study, experts
agreed that mycophenolate mofetil and cyclophospha-
mide are effective treatments for SSc-ILD.\(^ {7}\)

There is a lack of data from randomized controlled
trials on the efficacy and safety of immunosuppression
in the treatment of ILDs other than SSc-ILD, but several
clinical trials of immunomodulatory therapies in patients
with other autoimmune disease-related ILDs are ongo-
ing.\(^ {29}\) NPs should ensure that patients given immuno-
suppressant therapy are aware of its potential adverse reactions, such as an increased risk of infection, bone
marrow suppression, and gastrointestinal/liver toxicity, and of the requirement for regular monitoring of hepatic
function, renal function, and blood counts.\(^ {30}\) Adverse
reactions of glucocorticoids, such as weight gain and
sleep disturbance, can be bothersome and patients should
be advised on how to manage them.\(^ {31,32}\) Importantly,
treatment guidelines for IPF do not support chronic use
of immunosuppressants and recommend against use
of the combination of prednisone, azathioprine, and

\(N\)-acetylcysteine, following the PANTHER-IPF (Predni-
sone, Azathioprine, and \(N\)-Acetylcysteine: A Study That
Evaluates Response in IPF) trial, which demonstrated an
increased risk of hospitalization and death in patients re-
ceiving such therapy.\(^ {33,34}\) The value of corticosteroids and
other immunomodulatory therapies in treating chronic
hypersensitivity pneumonitis remains unclear and may
depend on the specific phenotype of the patient.\(^ {35,36}\)

**Antifibrotic therapies.** Nintedanib is a tyrosine
kinase inhibitor that inhibits processes involved in the
progression of pulmonary fibrosis such as the prolif-
eration and migration of fibroblasts and the deposition
of extracellular matrix components.\(^ {37}\) Nintedanib has
been approved by the FDA for the treatment of IPF
and chronic fibrosing ILDs with a progressive pheno-
type, as well as to decrease the rate of decline in FVC
in patients with SSc-ILD.\(^ {38}\) Randomized placebo-con-
trolled trials have shown that in patients with these
fibrosing ILDs, nintedanib reduces the rate at which
FVC declines.\(^ {39-43}\) The adverse reactions of nintedanib
are predominantly gastrointestinal events, particularly
diarrhea, which should be managed with hydration,
antidiarrheal medications such as loperamide, and, if
needed, by dose reduction or treatment interruption.\(^ {38}\) It should be
taken with food to reduce the risk of adverse reactions. Nintedanib may
also cause elevations in liver en-
zymes; liver function tests should be
conducted prior to treatment, at
regular intervals during the first 3 months of treat-
ment, and then periodically or as clinically indicated.\(^ {38}\)

As an inhibitor of the vascular endothelial growth
factor receptor, nintedanib may increase the risk of
bleeding. As such, nintedanib should be used with cau-
tion in patients receiving full-dose anticoagulation
therapy.\(^ {38}\) The bleeding events reported most frequently
in patients treated with nintedanib in clinical practice
are epistaxis, contusion, and hematochezia.\(^ {44}\) Arterial
thromboembolic events have been reported in patients
treated with nintedanib and caution is recommended
when using nintedanib in patients at higher cardiovas-
cular risk, including known coronary artery disease.\(^ {38}\)

The mechanism of action of pirfenidone has not
been established, but at high doses, it has been shown
to have antifibrotic effects, including a reduction in
the deposition of extracellular matrix components.\(^ {45}\)
Pirfenidone has been approved by the FDA for the treat-
ment of IPF, based on its ability to reduce the rate of
decline in FVC in these patients. In a placebo-controlled trial of pirfenidone in patients with unclassifiable ILD, analysis of the primary endpoint of change from baseline in the percent predicted value of FVC over 24 weeks, measured using daily home spirometry, was unable to be completed, but secondary endpoints, including change from baseline in the percent predicted value of FVC over 24 weeks based on in-clinic spirometry, suggested a benefit of the drug. There have been no randomized controlled trials of pirfenidone in patients with ILDs other than IPF and unclassifiable ILD. Adverse reactions associated with pirfenidone include gastrointestinal disorders, photosensitivity reactions, and rash, which may require dose reductions or treatment interruptions. Patients taking pirfenidone should be instructed to minimize exposure to sunlight and wear sunscreen. Liver function tests should be conducted prior to the initiation of therapy, monthly for the first 6 months, then every 3 months, and as clinically indicated.

Nonpharmacologic therapies. Patients with ILD have complex medical issues and their care requires a holistic and multidisciplinary approach. Exercise training/pulmonary rehabilitation programs can help to improve patients' symptoms and quality of life. Provision of supplemental oxygen may be beneficial for patients with severe resting hypoxemia, exertional desaturation, or severe dyspnea. However, when discussing oxygen therapy with patients, NPs should be mindful of the practical and psychosocial challenges that patients face in using supplemental oxygen. Education is vital to help patients understand their disease and set expectations regarding treatment and progression. Patients often turn to the internet to research their disease and are alarmed by what they find, much of which is inaccurate or misleading or not relevant to their specific diagnosis. NPs play a critical role in providing patients with accurate information and answering their questions. Patient support groups (live or virtual), such as those run by the Pulmonary Fibrosis Foundation, can be a great source of support for patients; NPs should provide information on these groups to their patients. Supportive/palliative care should not be limited to end of life but provided throughout the patient journey, alongside pharmacologic treatments.

Conclusions

Fibrosing ILDs have a variable clinical course. A proportion of patients with chronic fibrosing ILDs develop a progressive phenotype characterized by loss of lung function, worsening symptoms, and high mortality. An accurate diagnosis and regular monitoring to assess disease progression are vital to inform patient care and counseling. NPs have a key role to play within a multidisciplinary care team in helping patients understand their disease and its treatment, manage the adverse reactions of medication, and feel supported throughout the course of their disease.

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