doi.org/10.58931/cpct.2025.3138

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Essential Interstitial Lung Disease Management for the Primary Care Provider

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Abstract

Interstitial lung diseases (ILDs) encompass a diverse group of disorders characterized by inflammation and fibrosis of the lung parenchyma. Despite their classification as rare, increasing evidence suggests ILDs are more prevalent than previously thought. Patients often present with respiratory symptoms such as exertional dyspnea, persistent cough, and fatigue. However, asymptomatic patients with incidental findings on imaging (e.g., interstitial lung abnormalities) are also common. Diagnosis relies on high-resolution CT (HRCT), pulmonary function tests, and detailed clinical evaluation. Respirology consultation is important for comprehensive management. The evolving ILD nomenclature, including progressive pulmonary fibrosis, aids in disease characterization and treatment planning. Management strategies include corticosteroids and steroid-sparing agents for inflammatory subtypes, while antifibrotic therapies (nintedanib, pirfenidone) are used for fibrotic and progressive disease. Non-pharmacological interventions, including pulmonary rehabilitation, smoking cessation, and vaccination, are critical for improving patient outcomes. Primary care providers play a pivotal role in early disease recognition, facilitating diagnostic testing, managing comorbidities, and coordinating specialist care. This review highlights the importance of timely diagnosis, evolving classifications, and emerging therapies, offering a collaborative framework for optimizing ILD care and outcomes.

Introduction

Interstitial lung diseases (ILDs), or pulmonary fibrosis, are a heterogeneous group of disorders characterized by inflammation and/or fibrosis of the lung parenchyma. This umbrella term encompasses diseases with similar clinical, physiological, radiological, and pathological features. Although often considered to include

Terminology	Presentation	Imaging	Management
Interstitial lung abnormalities	Incidental finding on CT in someone not known to have ILD	Terms may include reticulation, fibrosis, lung distortion, ground glass opacities, traction bronchiectasis, honeycombing, non-emphysematous cysts	Clinical assessment PFTs Respirology referral (especially when fibrotic features are present i.e. traction bronchiectasis, honeycombing, or progression)
Fibrotic ILDs			
Idiopathic pulmonary fibrosis	Older age, cough and dyspnea on exertion	UIP pattern: subpleural, reticulation, traction bronchiectasis ± honeycombing	PFTs, HRCT, Respirology referral
Autoimmune-ILD (aka SARD-ILD)	Presentation may vary	UIP, NSIP, fHP, or OP pattern	Co-management with PCP, respirology, and rheumatology
Fibrotic hypersensitivity pneumonitis	Indolent with progressive cough and dyspnea; Exposure to organic antigen detected in 50% (mold, bacteria, birds, among others)	fHP pattern: upper and mid lung Peribronchovascular fibrosis, traction bronchiectasis, mosaic attenuation	PFTs, HRCT Respirology referral
Occupational ILD	Exposure history to asbestos, silica, beryllium, metal dust, among others	Varies by exposure	Antigen avoidance PFTs, HRCT Respirology referral ± WSIB
Unclassifiable ILD	Presentation varies, often cough and dyspnea	Fibrosis in indeterminate pattern	PFTs, HRCT Respirology referral
Non-fibrotic ILD			
Non-fibrotic hypersensitivity pneumonitis	May be an acute presentation following exposure to an organic antigen (mold, bacteria, birds, among others)	Ground glass opacities, air trapping, centrilobular nodules	May require inpatient admission vs urgent respirology referral
Organizing pneumonia	Cough, dyspnea, fever, similar to pneumonia Non-responsive to antibiotics	Migrating opacities, central ground glass opacity surrounded by consolidation ring (Atoll sign)	Corticosteroids with taper over 3+ months, steroid-sparing agent may be required for relapse Respirology referral
DIP/ RB-ILD	Smoking related disease, rarely associated with an autoimmune cause	Centrilobular nodules, ground glass opacities	Smoking cessation Respirology referral

Terminology	Presentation	Imaging	Management
ILD phenotypes			
Progressive pulmonary fibrosis	Progression of symptoms, PFTs ± fibrosis on CT	Varies by disease subtype, fibrotic features usually present	Respirology referral
Familial pulmonary fibrosis	Two or more family members affected	Varies, may not be a typical radiologic pattern	PFTs, HRCT Respirology referral
Combined pulmonary fibrosis and emphysema	Smoking history, may not have obstructed PFTs, markedly reduced DLCO	Upper lobe emphysema and lower lobe fibrosis	COPD management Respirology referral for consideration of antifibrotic therapy

Table 1. Common types of interstitial lung diseases and select phenotypes. Other rare forms of ILD not included: lymphocytic interstitial pneumonia, sarcoidosis, pleuroparenchymal fibroelastosis, lymphangioleiomyomatosis, post-COVID-19 fibrosis, drug-induced ILD, and pulmonary alveolar proteinosis; *courtesy of Amanda Grant-Orser, MBBCh, FRCPC*

Abbreviations: aka: also known as; **ILD:** interstitial lung disease; **CT:** computed tomography; **PFTs:** pulmonary function testing; **UIP:** usual interstitial pneumonia; **HRCT:** high-resolution CT; **SARD-ILD:** systemic autoimmune rheumatic disease – ILD; **NSIP:** nonspecific interstitial pneumonia; **fHP:** fibrotic hypersensitivity pneumonitis; **OP:** organizing pneumonia; **PCP:** primary care physician; **WSIB:** workplace safety and insurance board; **DIP:** desquamative interstitial pneumonia; **RB-ILD:** respiratory bronchiolitis ILD; **DLCO: d**iffusion capacity for carbon monoxide; **COPD:** chronic obstructive pulmonary disease

over 200 different types, this estimate is likely exaggerated, as classical classification systems recognize fewer subtypes.[1] ILD is considered a rare disease, but recent prevalence studies suggest it may be more common than previously thought, with rates ranging from 20 to 108 per 100,000 at risk Canadians.[2,3] ILDs are typically categorized as either idiopathic or secondary to another underlying condition or precipitant.[4] ILD nomenclature can be confusing and is frequently updated—for example, the recent shift from "acute and chronic hypersensitivity pneumonitis (HP)" to "non-fibrotic and fibrotic HP".[5] Keeping abreast with these evolving classifications can be challenging without actively following the latest literature. Regardless of the subtype, when fibrosis is present, indicated by computed tomography (CT) radiologic features of honeycombing and traction bronchiectasis, the process is irreversible, and often progressive. Due to the chronic and progressive nature of many ILD subtypes, involving a respirologist in patient care is encouraged. This review summarizes current ILD nomenclature, highlights when to suspect ILD, suggests initial investigations to consider, outlines how respirologists approach cases, and discusses current management options for patients.

When to Suspect ILD and Initial Investigations

Patients with ILD typically present in one of two ways: either they are symptomatic, or the disease is identified incidentally through imaging. Common symptoms include a dry or occasionally productive cough, shortness of breath (particularly with exertion), and fatigue.[6] Due to smoking being a shared risk factor with other diseases like chronic obstructive pulmonary disease (COPD) and coronary artery disease, misdiagnosis is common.[7-9] Additionally, comorbid respiratory diseases with similar symptoms can make an accurate diagnosis difficult. Since inhaler therapy is ineffective in ILD, the diagnosis should be considered in patients who do not respond to conventional treatments. ILD may be "unmasked" by viral infections, therefore, persistent respiratory symptoms following an upper respiratory tract infection warrant further evaluation.[10] Age is also important, as ILD typically presents after the age of 50. However, it may appear earlier in those with an autoimmune or drug-related presentation.[11,12]

Certain risk factors, such as family history, increase the likelihood of ILD. Individuals with a family history of ILD (defined as two or more

affected relatives within the same pedigree) have an almost 30% chance of abnormal CT findings.[13,14] Although screening is not currently recommended, clinicians should maintain a high index of suspicion in symptomatic patients with crackles during examination and who have a relevant family history. Any rheumatic disease may manifest with ILD. The autoimmune diseases most frequently affected by ILD include scleroderma (25-45%), myositis (30-80%), and rheumatoid arthritis (10-30%).[15,16]

Incidental radiologic findings, termed interstitial lung abnormalities (ILAs), are becoming more common due to the growing use of CT imaging. ILAs can be detected during lung cancer screenings, coronary CT scans, nodule follow-ups, or even in the lower slices of abdominal CT scans and upper slices of head and neck CT scans. Although ILAs should be considered a significant incidental finding, their description is often not included in the "impression" section of reports and therefore may be missed.[17,18] Terms such as 'subpleural reticulation', 'fibrosis', 'traction bronchiectasis', and 'honeycombing' are important to identify.[19] In such cases, a detailed respiratory history, risk factor assessment, physical examination, pulmonary function tests (PFTs), and referral to respirology are recommended. ILAs may represent undiagnosed ILD and require longitudinal follow-up.[20]

Physical examination is valuable, as velcro-like crackles at the lung bases are present in up to 90% of cases and can be easily auscultated in even mild fibrotic disease with excellent observer agreement.[21-23] Inspiratory squeaks can indicate small airways disease, which is a feature of hypersensitivity pneumonitis.[24] Digital clubbing may occur in up to 50% of patients.[25] While PFTs are an essential test, they can be normal in mild ILD, making them unreliable for ruling out the disease.[26,27] Laboratory testing is nonspecific for ILD but can help in identifying autoimmune diseases through serologies such as rheumatoid factor, anti-citrullinated peptide, and antinuclear antibody, while extractable nuclear antigen and myositis serology testing should be considered on a case-by-case basis.[28,29]

The gold standard for diagnosing ILD is a high-resolution CT (HRCT) performed with an "ILD protocol," which includes inspiratory, expiratory, and prone imaging.[28] Chest radiographs are not recommended for ILD screening. HRCT should be considered for high-risk patients, including those with symptoms, a family history, autoimmune comorbidities, crackles on examination, restrictive PFTs, or abnormal chest radiographs suggestive of ILD. Patients with ILD on CT at the time of referral are often prioritized for respirology review, which significantly expedites their care.[7]

How Respirologists Diagnose ILD

Patients with ILD undergo a comprehensive evaluation upon referral to respirology, including a review of symptoms, timelines, precipitating events, and risk factors. Some practitioners may also use a questionnaire to identify environmental exposures that could contribute to the disease.[30] Baseline investigations might include liver function tests, infection screening, and autoimmune serologies to assess for comorbidities or prepare for medication initiation. PFTs, particularly forced vital capacity and diffusing capacity for carbon monoxide (DLCO), are useful for assessing disease severity, predicting mortality, and monitoring disease progression.[31,32] A six-minute walk test is helpful for evaluating exertional hypoxia, which may qualify some patients for home oxygen based on provincial eligibility criteria.[33] HRCT is essential for confirming an ILD diagnosis, and ideally, images should be reviewed by a thoracic radiologist. The radiologic pattern provides critical diagnostic insights into the ILD subtype. For instance, a usual interstitial pneumonia (UIP) pattern is observed in idiopathic pulmonary fibrosis (IPF), while nonspecific interstitial pneumonia (NSIP) may indicate autoimmune disease-related ILD, drug-induced ILD, or other conditions.[16,31,34]

Bronchoscopy is not routinely performed but may be used to rule out infection, assess for diffuse alveolar hemorrhage, or evaluate inflammatory markers (e.g., lymphocytosis on cell count and differential).[5,28] Transbronchial biopsy is not recommended to diagnose most forms of ILD, although may be useful in some clinical scenarios. Surgical lung biopsy is rarely performed and should only be considered after a case discussion in an ILD multidisciplinary discussion (MDD).[28] MDDs are a standard component of an ILD diagnostic workup, providing input from a team of experts.[35,36] While most ILD cases can be managed by general respirology, access to MDDs enhances diagnostic accuracy.[35] Virtual referrals to ILD programs, often located in tertiary care centres, are available in many regions to support community respirologists. These programs typically include specialized ILD physicians,

fellows, and allied health professionals. In addition, many community respirologists have undertaken dedicated ILD training.[37]

Patients with ILD are subsequently classified into specific subtypes[4] **(Table 1)**. Beyond traditional classifications, phenotypes are increasingly used to guide treatment. The term "progressive pulmonary fibrosis" (PPF) describes patients with worsening symptoms, PFTs, and/or imaging findings, who may benefit from antifibrotic therapies.[31] Familial pulmonary fibrosis is used to describe patients with a family history of the disease, who often experience a more aggressive disease course.[38] Combined pulmonary fibrosis and emphysema (CPFE) syndrome describes the simultaneous occurrence of COPD and ILD.[39] Novel endotyping techniques, including telomere length testing, genetic analyses, and predictive biomarkers, hold promise for ILD management but are currently limited to tertiary ILD programs or research settings.[40-42]

Current and Emerging Therapies for ILD

Management of ILD involves both pharmacological and non-pharmacological approaches. For inflammatory subtypes, such as autoimmune disease-related ILD, non-fibrotic hypersensitivity pneumonitis, or drug-induced ILD, corticosteroids are often initiated, followed by steroid-sparing agents like mycophenolate or azathioprine.[43,44] Fibrotic and progressive disease subtypes, including IPF and PPF, are treated with antifibrotic medications.[31] Currently, two antifibrotics are approved for use to slow the progression of ILD: nintedanib (Ofev), indicated for IPF and PPF, and pirfenidone (Esbriet), approved for IPF.[45-47] Both require liver function monitoring and may cause gastrointestinal side effects, limiting their tolerability. Symptom management, such as treating cough with over-the-counter suppressants, liquid codeine, or low-dose morphine, is also common.[48] Proton pump inhibitors are only recommended for those with

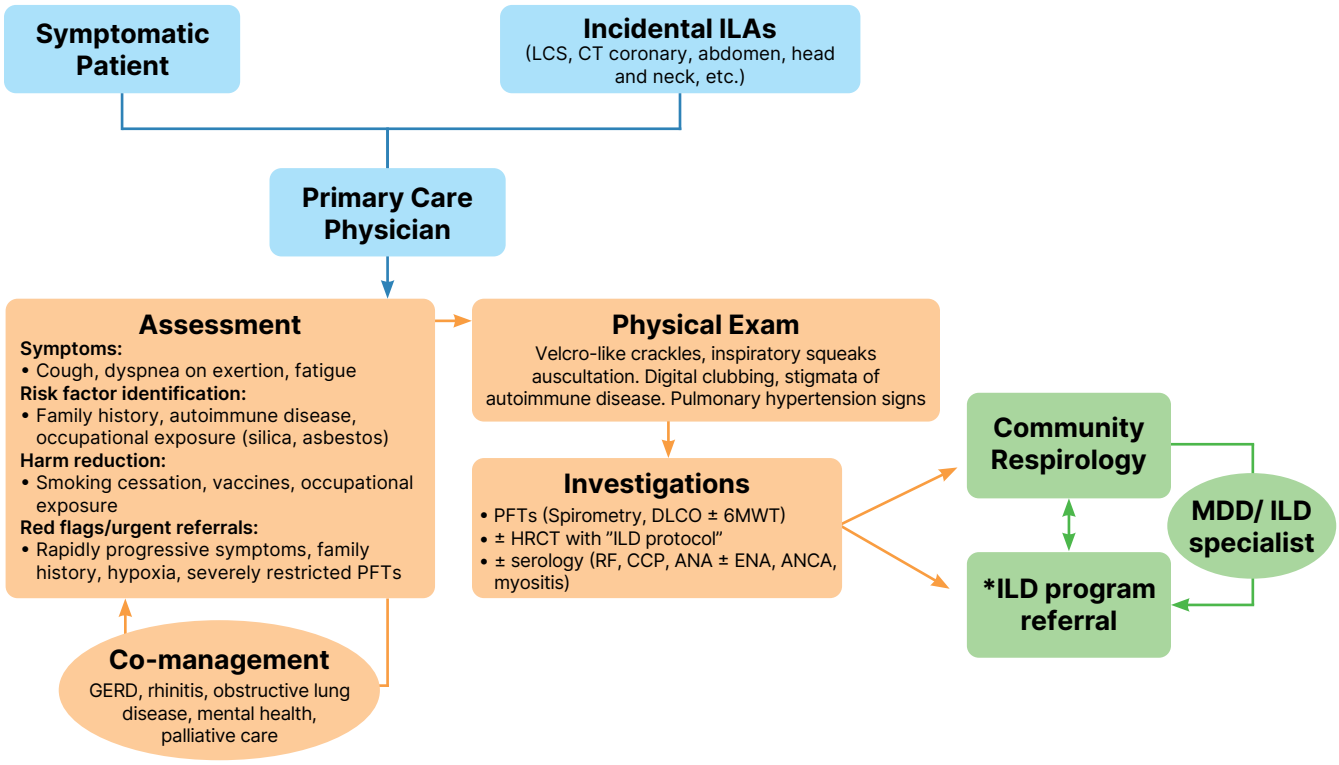

Figure 1. Suggested algorithm for ILD work up and referral; *courtesy of Amanda Grant-Orser, MBBCh, FRCPC*

*Referral patterns and access to ILD programs may vary by region

Abbreviations: ILAs: interstitial lung abnormalities; **PFTs:** pulmonary function testing; **DLCO:** diffusion capacity of carbon monoxide; **6MWT:** six-minute walk test; **HRCT:** high-resolution computed tomography; **ILD:** interstitial lung disease; **RF:** rheumatoid factory; **CCP:** Anti-citrullinated protein; **ANA:** antinuclear antibody; **ENA:** extractable nuclear antigen; **ANCA:** antineutrophil cytoplasmic antiody; **MDD:** multidisciplinary discussion; **GERD:** gastroesophageal reflux disease.

symptomatic reflux disease.[49] For advanced or progressive disease, lung transplantation may be considered.

Non-pharmacological management focuses on risk factor modification. Patients with hypersensitivity pneumonitis are advised to avoid exposure to antigens.[50] Smoking cessation is essential as it may slow the progression of ILD and mitigate the synergistic risk of developing lung cancer associated with smoking and ILD.[51] Vaccinations, including those for influenza, COVID-19, and pneumococcal infections are strongly recommended. Respiratory syncytial virus (RSV) vaccination should also be considered. While home oxygen therapy does not improve survival, it enhances quality of life.[52] Pulmonary rehabilitation and early referral to palliative care are similarly beneficial and encouraged.[53-55] Patient support groups are available through the Canadian Pulmonary Fibrosis Foundation (cpff.ca).

How Can Primary Care Providers Co-Manage ILD?

Primary care is the cornerstone of Canadian healthcare and often the first point of contact for patients with ILD **(Figure 1)**. Early suspicion by primary care physicians (PCPs) is crucial for improving care, enabling timely testing (e.g., PFTs, HRCT) and referral to respirology. PCPs play a key role in addressing risk factors, including promoting vaccination, smoking cessation, and reducing occupational exposures. They also frequently manage comorbid conditions such as gastroesophageal reflux disease (GERD), postnasal drip, and asthma. Additionally, PCPs assess mental health, provide palliative care support, or facilitate access to such services, which are essential in ILD. Advocating for patients, coordinating with specialists, and fostering clear communication significantly enhances patient care and outcomes.

Conclusion

In summary, early recognition and appropriate testing for ILD are critical responsibilities for PCPs. By maintaining a high index of suspicion, identifying risk factors, initiating key diagnostic tests such as PFTs and HRCT, noticing incidental radiologic findings and facilitating timely referrals to respirology, PCPs can significantly impact patient outcomes. Additionally, addressing comorbidities, advocating for lifestyle modifications, and providing palliative care when needed ensure comprehensive, collaborative care for patients with ILD.

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Financial Disclosures

A.G.O.: Honoraria: Boehringer Ingelheim.

Resources

Patient information and support groups
Canadian pulmonary fibrosis foundation
www.cpff.ca

Canadian ILD respirologists
Find an ILD Respirologist in Canada 2024
on cpff.ca

Canadian Thoracic Society Guidelines and Position Statements
CTS guideline library
cts-sct.ca/guideline-library

American Thoracic Society Guidelines
ATS Official Documents, Interstitial Lung Disease
www.thoracic.org/statements/insterstitial-lung-disease.php

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