Improving care in progressive fibrosing ILD: the need for multidisciplinary guidelines informed by key stakeholders

Backgrounder

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Working document prepared in support of policy discussions related to PF-ILD in Canada.

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BACKGROUND

Providing optimal care (diagnosis, treatment, support and long-term monitoring) for patients with progressive fibrosing interstitial lung disease (PF-ILD) represent substantial time and healthcare resources and a potentially complex patient journey. In a global (US, Japan, Germany, France, Italy, Spain, and UK) survey of 486 physicians (243 respirologists, 203 rheumatologists, 40 internists), it was estimated that PF-ILD due to scleroderma (SSc-ILD) is typically diagnosed 8.6 to 11.5 months after symptoms develop.[1] Extended time to diagnosis is a concern for patients awaiting appropriate care and care providers who need to recognize and manage early complications from scleroderma.

Patient journeys in PF-ILD associated with other complex conditions like rheumatoid arthritis (RA) follow a similar course, with frustration over disability, negative impacts on personal relationships, reduced productivity and the need for education featuring highly as unmet needs. Similarly observed lag times to diagnosis and treatment have also been reported in RA.[2]

The Patient Journey and the need for a multidisciplinary approach to evaluation

The patient journey is a time-consuming and difficult one, not necessarily because of a lack of resources or because it represents a low health system priority. Rather, appropriate diagnosis requires multiple steps involving multiple specialities and services from radiologists, primary care providers, rheumatologists, and other specialists. The need for follow-up, referral, tests and awaiting definitive diagnoses feature prominently. A patient pathway based on this research[40] is depicted in **Figure 1**.

Existing clinical guidance

The Canadian Thoracic Society, as part of its educational mandate and need to provide respiratory specialists with tools to optimize care, regularly develops guidelines and position statements that address diagnosis and therapeutic management across a number of areas, including asthma, chronic obstructive pulmonary disease, home mechanical ventilation, sleep disorders, pediatric conditions and pulmonary vascular diseases. In 2017 and 2018, the Canadian Thoracic Society Clinical Assembly on Interstitial Lung Disease (CTS-AILD) released two consensus position statements separately addressing the evaluation and care of patients.[3]

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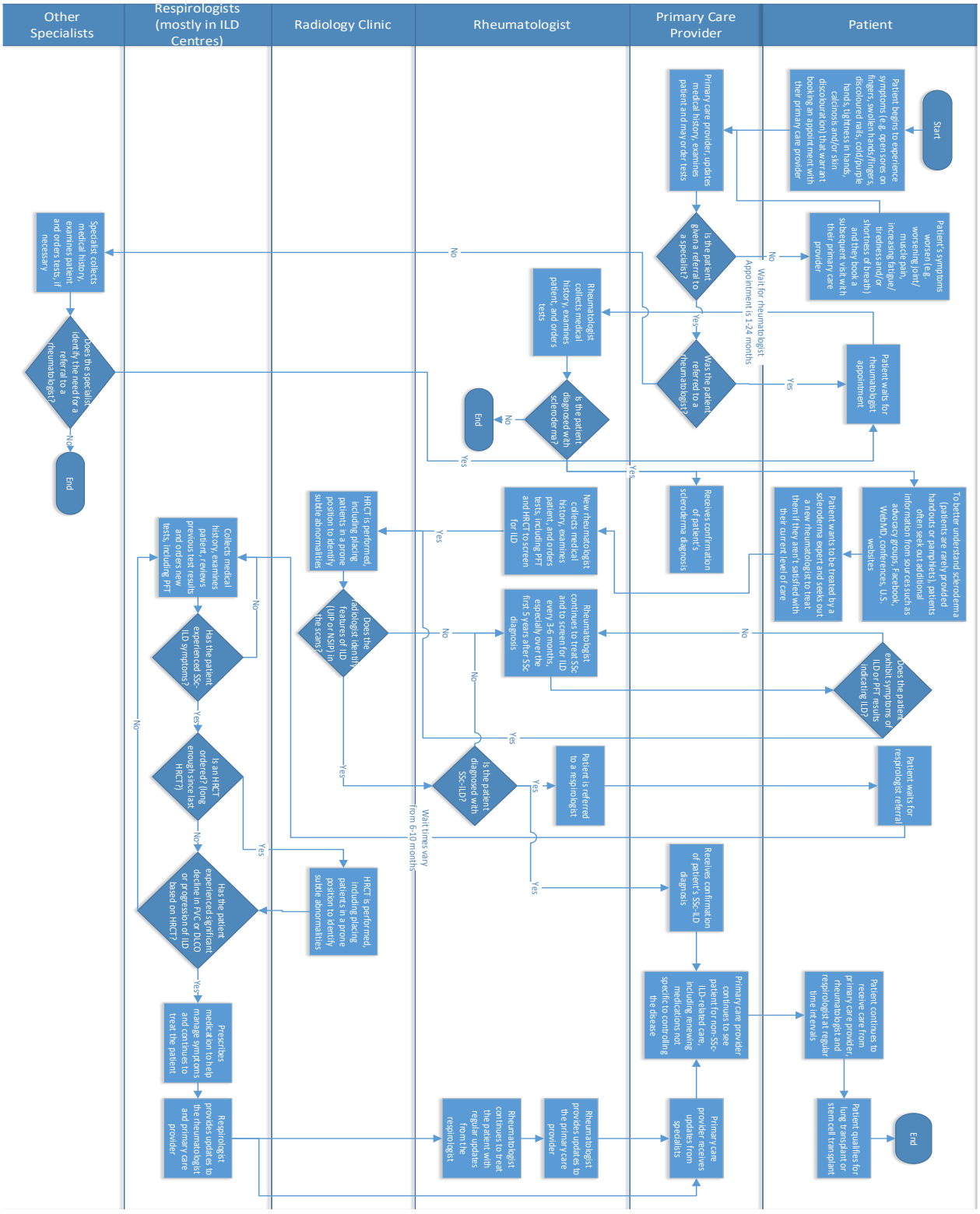
[1] "Canadian Rheumatology Association Meeting Fairmont The Queen Elizabeth Montreal, Quebec, Canada February 27 – March 2, 2019," *The Journal of Rheumatology* 46, no. 7 (July 1, 2019): 757–865, https://doi.org/10.3899/jrheum.190333.#33 Non-IPF Progressive Fibrosing Interstitial Lung Disease (PF-ILD): The Patient Journey

[2] Alaa S. Barhamain et al., "The Journey of Rheumatoid Arthritis Patients: A Review of Reported Lag Times from the Onset of Symptoms," *Open Access Rheumatology: Research and Reviews* 9 (2017): 139–50, https://doi.org/10.2147/OARRR.S138830; Javier E. Rosa et al., "Rheumatoid Arthritis Patient's Journey: Delay in Diagnosis and Treatment," *Journal of Clinical Rheumatology: Practical Reports on Rheumatic & Musculoskeletal Diseases*, October 11, 2019, https://doi.org/10.1097/RHU.0000000000001196.

[3] Kerri A. Johannson et al., "Evaluation of Patients with Fibrotic Interstitial Lung Disease: A Canadian Thoracic Society Position Statement," *Canadian Journal of Respiratory, Critical Care, and Sleep Medicine* 1, no. 3 (July 3, 2017): 133–41, https://doi.org/10.1080/24745332.2017.1359056; Deborah Assayag et al., "Comprehensive Management of Fibrotic Interstitial Lung Diseases: A Canadian Thoracic Society Position Statement," *Canadian Journal of Respiratory, Critical Care, and Sleep Medicine* 2, no. 4 (October 2, 2018): 234–43, https://doi.org/10.1080/24745332.2018.1503456.

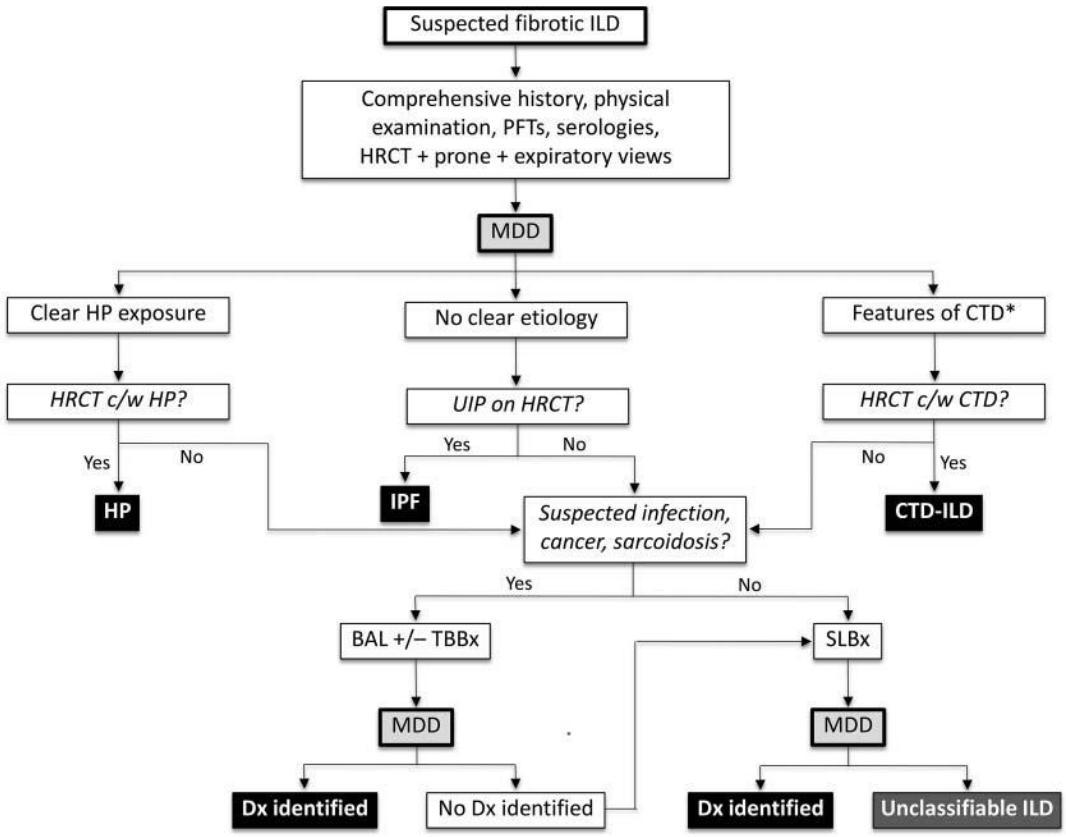
Figure 1 Multidisciplinary care and the patient journey



In its creation of key messages for the evaluation of patients, the positions paper notes that "ILD patients should be reviewed in a multidisciplinary discussion (MDD) whenever possible before initiating disease-specific pharmacotherapies". This recommendation was similarly made in an official 2018 clinical practice guideline jointly developed and endorsed by The American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Latin American Thoracic Society (joint ATS/ERS/JRS/ALAT or International Panel).[4]

MDD is defined by the International Panel as "interaction between a pulmonologist (and rheumatologist on a case-by-case basis), radiologist, and pathologist."[5] While both Canadian and International guidelines recognize interactions could be conducted in person or virtually, e.g., telephone, Internet/e-mail, text, and/or reading interpreted reports, the joint CTS-AILD guidelines encouraged face-to-face or voice-to-voice MDDs when formal clinical reports of the interpretation by experts in different disciplines are in discordance. The algorithm proposed by the CTS-AILD is shown in **Figure 2**.

Figure 2: Approach to the evaluation of fibrotic interstitial lung disease.[6]



* Clinical or serological features of CTD

[4] Ganesh Raghu et al., "Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline," *American Journal of Respiratory and Critical Care Medicine* 198, no. 5 (September 2018): e44–68, https://doi.org/10.1164/rccm.201807-1255st.

[5] Raghu et al.

[6] Johannson et al., "Evaluation of Patients with Fibrotic Interstitial Lung Disease."

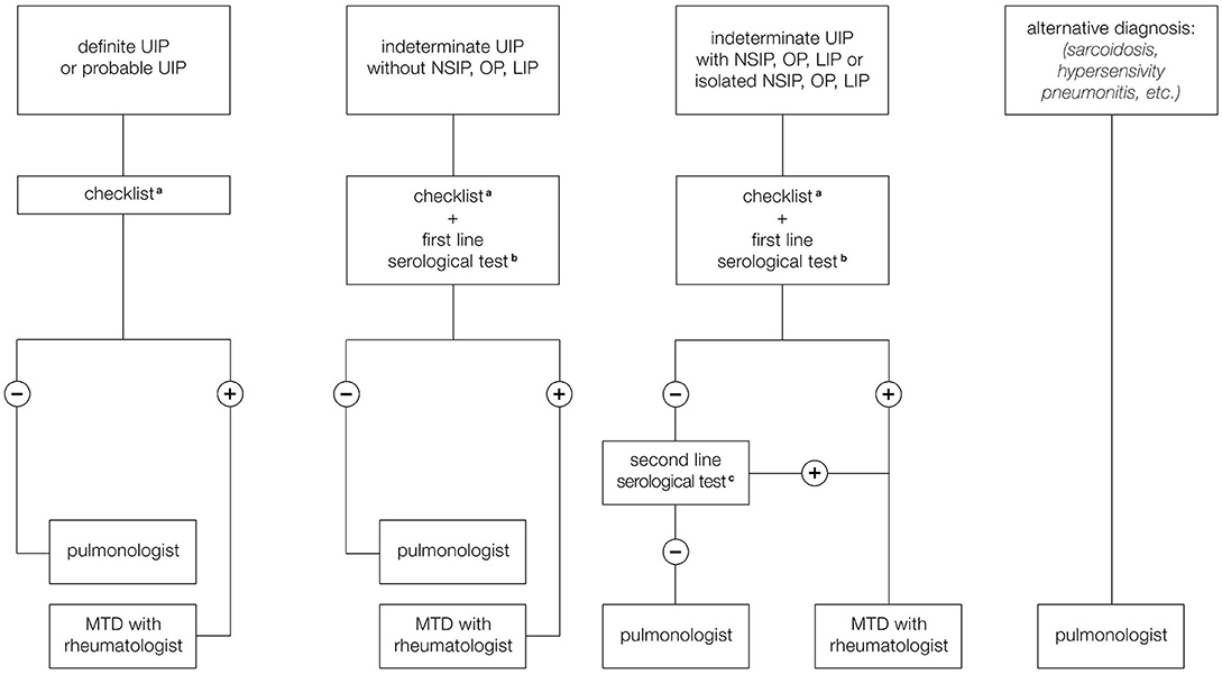
Abbreviations: CTD, connective tissue disease; HRCT, high resolution computed tomography; ILD, interstitial lung disease; IPF, idiopathic pulmonary fibrosis; MDD, multidisciplinary discussion; UIP, usual interstitial pneumonia.

Evidence to support the use of a multidisciplinary approach

Despite a positive recommendation for an MDD approach by both Canadian and international panels, each clinical practice guidelines acknowledged a low level of evidence to support MDD, even after a systematic review of all available evidence. While some guidance is provided for incorporating an MDD into practice, an optimal algorithm, or precise exact care pathway defining when and how different disciplines should be engaged is not defined.

Since these 2018 recommendations, an updated systematic review has been published.[7] The authors qualitatively synthesized 29 research reports examining the use of an MDD approach in the evaluation of ILD patients. While acknowledging existing recommendations, the authors also attempt to define an optimal MDD approach to evaluation based on available evidence. Their proposed care pathway is shown in **Figure 3**.

Figure 3: Proposal for a multidisciplinary team (MDT) involving the rheumatologist.



(a) Checklist regarding signs and symptoms compatible with CTD or arthritis. (b) First line serological test: RF, ACPA, ANA, CPK. (c) Second line serological test: Anti-ds DNA, Anti-Ro (SS-A), Anti-La (SS-B), Anti-ribonucleoprotein, Anti-topoisomerase (Scl-70) Anti-tRNA synthetase, Anti-PM-Scl, Anti-MDA5.

Evidence to support the use of an MDD approach was also identified for the routine management of patients diagnosed with ILD. Some evidence was identified in the systematic review suggested patients

[7] Federica Furini et al., "The Role of the Multidisciplinary Evaluation of Interstitial Lung Diseases: Systematic Literature Review of the Current Evidence and Future Perspectives," *Frontiers in Medicine* 6 (2019): 246, https://doi.org/10.3389/fmed.2019.00246.

evaluated by MDD experienced greater satisfaction and more participation in their care path[8] and that a multidisciplinary approach in palliative care involving the participation of ILD expert could have a positive impact on patients in terms of reduced number of emergency visits and hospital admissions.[9]

Other recent publications shed some light on how MDD is being conducted internationally. A global survey of 457 unique centres across 64 countries revealed 350 (76.6%) centres holding formal meetings, with the majority holding face-to-face MDT meetings (80%), for a minimum of 30 min (93%), and discussed diagnosis (96.9%) and patient management (94.9%) at the meetings. ILD academic centres reported a higher ILD caseload, held more formal MDT meetings, and were more likely to include histopathology and rheumatology specialists in their diagnostic team. Of the centres holding MDT meetings, 5.5% routinely discussed all new cases at such meetings.[10] Specific descriptions of the role of MDD in ILD services in Singapore[11] and Canada (Vancouver)[12] have also been published.

[8] Shin Ok Jeong et al., "Effects of Patient Satisfaction and Confidence on the Success of Treatment of Combined Rheumatic Disease and Interstitial Lung Disease in a Multidisciplinary Outpatient Clinic," *International Journal of Rheumatic Diseases* 21, no. 8 (2018): 1600–1608, https://doi.org/10.1111/1756-185X.13331.

[9] Meena Kalluri et al., "Beyond Idiopathic Pulmonary Fibrosis Diagnosis: Multidisciplinary Care With an Early Integrated Palliative Approach Is Associated With a Decrease in Acute Care Utilization and Hospital Deaths," *Journal of Pain and Symptom Management* 55, no. 2 (February 1, 2018): 420–26, https://doi.org/10.1016/j.jpainsymman.2017.10.016.

[10] Luca Richeldi et al., "The Characterisation of Interstitial Lung Disease Multidisciplinary Team Meetings: A Global Study," *ERJ Open Research* 5, no. 2 (April 2019), https://doi.org/10.1183/23120541.00209-2018.

[11] Gt Chai et al., "Impact of an Interstitial Lung Disease Service in the Diagnosis and Management of Interstitial Lung Disease in Singapore," *Singapore Medical Journal*, July 11, 2019, https://doi.org/10.11622/smedj.2019069.

[12] Japnam S. Grewal et al., "Role of a Regional Multidisciplinary Conference in the Diagnosis of Interstitial Lung Disease," *Annals of the American Thoracic Society* 16, no. 4 (April 2019): 455–62, https://doi.org/10.1513/AnnalsATS.201811-794OC.

Policy Options and Next Steps for Canada

In creating guidance for Canada, the CTS-AILD was explicit that the views and preferences of patients with PF-ILD were not considered and would be sought in future position statements. They also emphasized their desire to partner with other organizations and stakeholders to support the implementation of key messages. While updated position statements are currently in development, they are not expected to specify an approach to multidisciplinary discussion that incorporates the views of patients or rheumatologists.[13] As such, logical next steps might be to an define optimal care pathway that considers the patient journey and preferences and values of Canadian patients and participation of other specialists. This might be accomplished by the following steps:

1. Characterize the referral pathways currently being developed or used for the evaluation of PF-ILD patients through further literature review and nationwide survey.
2. Identify key opinion leaders in rheumatology (beyond scleroderma specialists) with an interest in defining optimal care pathways for patients with PF-ILD of autoimmune etiology.
3. Gather qualitative feedback from patients in regards to what referral pathways most align with patient values.
4. Based on the release of upcoming CTS evaluation and management guidelines advocating for multidisciplinary discussion, and working with these stakeholders, identify which aspects are most desirable and create further statements about what else is needed (i.e., specialized training, availability of specialists, novel platforms (such as Ontario e-consult) to support optimized referral.
5. Identify processes that are sustainable within the Canadian health system, with optimized efficiency and outcomes for patients.
6. Characterize the epidemiology of PF-ILD in Canada to understand how geographic barriers (i.e. urban vs rural status) impact the patient journey to diagnosis and treatment.

[13] Personal communication, Dr. Martin Kolb, 13-Jan-2019