INTERSTITIAL LUNG DISEASES (ILD) PROGRAM

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ILD PROGRAM

History and Overview

The ILD Program originated through the creation of an ILD Clinic in May, 2013 by Dr. Onofre Moran with the approval and support of Dr. Michael Fitzpatrick, Head of the Division of Respirology and Critical Care Medicine at that time. Before the ILD program was created, nothing like this existed, even though there was the need for it.

The ILD Clinic was created to attend the demands of a specialty clinic for patients with interstitial lung diseases, who, for their diagnosis and management, frequently require the participation of several specialties.

In 2014, the ILD Clinic rapidly evolved into an Interstitial Lung Disease Program that has included:

- ILD Clinics
- ILD Nurse
- ILD Multidisciplinary Rounds
- ILD Fellowship Program
- ILD Research Program
- ILD Program Collaborators

ILD Clinics

Before the ILD Clinic was implemented in May 2013, Dr. Moran had several meetings with various specialists who kindly agreed to collaborate and develop the standardized protocols currently used at the ILD Clinic. Dr. Moran also attended the ILD Program at the University of Toronto to learn the approach and procedures used by Dr. Shane Shapera, Director of their ILD Program, that he learned at the ILD Program in the University of California in San Francisco. Also, attended the Interstitial Lung Diseases (ILD) Programs at the Center of Excellence for ILD with Dr. Jurgen Behr in Munich, in Germany.
Dr. Moran went to Italy (with Dr. Venerino Poletti) and to Israel (with Dr. Mordechai Kramer) to train in transbronchial lung cryobiopsies (TBLC) and started the TBLC program at Kingston General Hospital in October 2018. During his rotation in Italy he also participated the ILD multidisciplinary discussions.

Dr. Kirsten Nesset did her training in ILD in McMaster and joined the Division of Respiratory and Sleep medicine and our ILD Program in the fall of 2024.

The ILD clinics are currently held at Hotel Dieu Hospital. Dr. Moran holds clinics from 8:30 am to 1 pm on Thursdays, and from 1 pm to 6 pm on Mondays and Tuesdays (the later combined with general Respirology clinic). Dr. Nesset holds ILD clinics from 8:30am to 1pm on Mondays and from 1pm to 5pm on Thursdays.

All diagnostic and management protocols and procedures used in our ILD clinic and Program are in compliance with the Guidelines for the Diagnosis and Management of Idiopathic Pulmonary Fibrosis (IPF), published by the American Thoracic Society (ATS), European Respiratory Society (ERS), Japanese Respiratory Society (JRS), and Asociacion Latino Americana de Torax (ALAT)[1-4] as well as International Guidelines for the Diagnosis of Idiopathic Interstitial Lung Diseases,[5] Hypersensitivity Pneumonitis[6,7] and the Guidelines on Bronchoalveolar Lavage (BAL) in ILD[8].

Following current ILD/IPF Guidelines and with collaboration and input from our colleagues from Chest Radiology, Rheumatology, Thoracic Surgery, Lung pathology, Hematopathology, Palliative Care and Pulmonary Rehabilitation, several protocols for patients' assessments and referrals were created and implemented and are re-assessed and updated usually on annual basis (see Appendices 1-5).

All patients attending the ILD Clinic undergo a protocol (see Appendix 1) with the following standardized assessments to properly investigate the most common groups of known causes of ILD: Hypersensitivity Pneumonitis, Connective Tissue Diseases (CTD), and drugs, as per Guidelines[1]:

- Questionnaires:
  - ILD Environmental and Drug Exposures Assessment Questionnaire. Which includes the American College of Chest Physicians (ACCP) and the "white paper" on Hypersensitivity pneumonitis questionnaires combined and modified (shown in Appendix 2)[9,10]
  -King's Brief ILD Questionnaire (see Appendix 3).
  -Connective Tissue Diseases Screening Questionnaire, work up and Rheumatology referral criteria (see Appendix 4).
- High Resolution Chest CT (HRCT) protocols with routine inspiratory, expiratory, and prone views and interpretation recommendations as per current Guidelines by Chest Radiologists (see Appendix 5).
- Pulmonary Function tests (PFTs), and 6-minute walk test (6MWT).
- Laboratory work-up for CTD and Vasculitis.

As per our ILD Protocol, patients with either clinical or laboratory abnormalities suggestive of a connective tissue disease are referred to Dr. Clements-Baker, Rheumatologist, who assesses our ILD patients (usually within 2-4 weeks) to determine if patients have a CTD that could explain their ILD.

Patients who have –as per the American College of Chest Physicians Questionnaire- environmental exposures associated with hypersensitivity pneumonitis or have exposure to drugs reported to cause ILD, are routinely offered bronchoscopy with bronchoalveolar lavage +/- transbronchial biopsies or cryobiopsies.

The bronchoalveolar lavage (BAL) procedure, transportation, sample processing, and analysis are performed as per the BAL in ILD Guidelines[8]. The BAL cell differential, essential in the diagnostic approach to ILD, are reported by Dr. David Good, as per the guidelines. Dr. Good also kindly agreed to "read" and report all of our BAL samples, and to collaborate with the academic ILD program by providing training sessions to our ILD Fellows on the processing and analysis of BAL samples.
We have weekly sessions to assess BAL results and plan the next steps based on those results.

Patients who have an indeterminate to UIP pattern on high resolution chest CT, when appropriate, are offered lung biopsy, either through TBLC or video assisted thoracoscopic surgery (VATS) and are discussed in our multidisciplinary ILD Rounds using a standardized approach as per current guidelines (see ILD Multidisciplinary Rounds below).

Once the diagnosis is established, if anti-fibrotic treatment is required, patients are re-scheduled in clinic or through a phone consult to discuss the pros and cons of treatment, as well as the need to refer them to:

- Dyspnea Clinic to be assessed and managed by our Palliative Care Physicians who have kindly agreed to collaborate with our ILD clinic and to manage the dyspnea in our patients. An example of the initial approach to manage dyspnea in our patients is shown in the Appendix 6.

- Pulmonary Rehabilitation Program, with Dr. Alberto Neder, to assess suitability for enrollment in the rehabilitation program he supervises at Providence Care Hospital. Referral criteria are shown in the Appendix 7.
-Lung Transplant Program in Toronto: eligible patients diagnosed with IPF or with another ILD, if advanced or progressive despite treatment, are referred to the lung transplant program in Toronto.

After diagnosis and treatment are established, our ILD patients are followed in our ILD clinic every 3-6 months -depending on the severity of their disease on presentation and the progression over time-, with: HRCT –as per protocol established with our Chest Radiologists, who read all the ILD patients' HRCTs-, full pulmonary function tests, 6-minute walk tests, and blood work if clinically indicated.

ILD Nurse

An ILD Nurse –Mrs. Lynda McCarthy- was hired in January 2015 to work at the ILD Clinic; since her retirement in 2022, Mrs. Carla Paredes was hired as our ILD Nurse, and was our ILD nurse until March 2025.

Our ILD nurse has several roles:
Before every appointment our ILD nurse ensures that patients have the appropriate chest CT, pulmonary function tests, and blood work; and, if there is missing information, she tries to schedule any missing tests as soon as possible so that the required information is available for the respirologists to make the proper decisions during the patient's clinic appointment.

Upon arrival of patients to clinic our ILD nurse collects the information from questionnaires mailed to patients before their appointment, checks all the results of the tests ordered, obtains the list of all drugs patients are on and ascertains their possible association with interstitial lung diseases via the website www.pneumotox.com. Then, she transfers all the clinically relevant information from the questionnaires, PFTs, 6-minute walk test, and bloodwork into our ILD Data Collection Form (see Appendix 8), which we use routinely in the assessment of patients in the ILD clinic.

She also transfers all the information from clinical questionnaires, blood work, lung function testing, imaging studies, bronchoscopy (BAL cell count and differential), and histopathological results, including the diagnosis achieved at the multidisciplinary ILD rounds, into an electronic database to be used in research studies.

After the first clinic visit, our ILD nurse provides all our ILD patients with the "ILD Clinic Patient Recommendations" (shown in Appendix 9) and written anti-gastroesophageal reflux disease measures. The patient recommendations address environmental exposures of risk that patients should avoid, as well as the monitoring and proper titration of their oxygen, for patients who require supplementary oxygen.

If bronchoscopy is required (which is the case in most patients), our ILD nurse arranges the appointment and provides patients with verbal and written information of the preparation required for the procedure, including modification to their medications before the procedure (Diabetic drugs, anticoagulants, antiplatelets, etc. as appropriate). She also ensures patients get the proper testing before bronchoscopy (coagulation tests and electrocardiogram) when applicable.

Our ILD Nurse routinely instructs our ILD patients in clinic about the proper use of a personal pulse oximeter, when appropriate (most cases).

The ILD Nurse also participates in the ILD Multidisciplinary Rounds and helps set up the follow-up appointments and/or starting patients on anti-fibrotic treatment, according to the diagnosis and plans achieved during the ILD rounds.

ILD Multidisciplinary Rounds

The ILD multidisciplinary rounds were created in July 2013 to present and discuss patients in whom surgical biopsy was required for the diagnosis of their ILD –as per ATS/ERS/JRS/ALAT and other pertinent Guidelines (as shown in Appendix 10).

The following disciplines collaborate with patients presented in the ILD rounds:

- Chest Radiology: Dr. Rob Dhillon and Dr. Dominique DaBreo, who read all the HRCTs on our ILD patients and one of them discusses the radiological findings at the ILD rounds.
- Lung Pathology: Dr. Alexander Boag, who describes the histopathological findings of the cases presented in the rounds.
- Respirology: Dr. Onofre Moran attends and coordinates the ILD rounds as well as the differential and final diagnostic and management plans of patients presented in rounds. Dr. Kirsten Nesset is an ILD specialist who since being hired in the fall of 2024, routinely attends our ILD Rounds, and provides her input and expertise in the diagnosis and management plans of patients presented in rounds. All other Respirologists in the Division of Respirology at Queen's University are also invited to present cases.
- ILD Fellows, Respirology Fellows and House Staff rotating in Respirology attend the ILD rounds and are responsible to present the cases following a standardized Power Point template.
- ILD Nurse: Helps coordinating the follow-up and management plans of patients after rounds.
- Thoracic Surgery: Dr. Kenneth Reid, Dr. Wiley Chung and Dr. Andrew Giles are our Thoracic Surgeons, who perform the video assisted surgical lung biopsies when required, as per the "Guidelines" recommendations[1].

The ILD rounds usually occur monthly and Dr. Moran creates the rounds schedule quarterly. To date, more than 200 ILD cases have been presented and discussed in the ILD Multidisciplinary Rounds.

The ILD Fellows and/or Respirology Fellows present the cases using a standard template that includes the clinical, functional, laboratorial, and bronchoscopic results of the case. Thereafter, the HRCTs are discussed by the chest radiologist. Then, Dr. Boag (our Lung Pathologist) discusses the histopathological findings from the Transbronchial Cryobiopsies or VATS biopsies. Finally, based on the diagnostic classifications recommended by the Guidelines[2-4,6,7,11] a final diagnosis is achieved in common agreement. Dr. Moran dictates a note that is uploaded into the patients' electronic medical records with each of the following diagnoses: Radiological, Pathological and Multidisciplinary diagnosis; as well as follow-up and management plans, as per the Guidelines. Our ILD nurse, who attends the ILD rounds, helps coordinate the implementation of the diagnostic/management plans. Lunch is routinely provided in ILD rounds.

ILD Fellowship Program

The ILD Fellowship program was developed by Dr. Onofre Moran in November 2013 and approved by the Postgraduate Medical Education Office of Queen's University in 2014. The first ILD Fellow –Dr. Muhannad Hawari- was accepted on July 1st, 2014. Since its inception, the following ILD Fellows have graduated from our program:

-2014-15 Dr. Muhannad Hawari
-2016-17 Dr. Sami Alyami
-2017-18 Dr. Sharina Aldhaheri and Dr. Sami Alyami (ILD Research Fellow)
-2018-19 Dr. Bader Alharthi and Dr. Sharina Aldhaheri (ILD Research Fellow)
-2019-20 Dr. Mohamed Khalil
-2020-21 Dr. Mohamed Khalil (ILD Research Fellow)
-2022-23 Dr. Salem Alqahtani and Dr. Ahmad Al-Jarallah
-2023-24 Dr. Alejandra Coppola-Lamas and Dr. Elham Alrubai
-2024-25 Dr. Hend Aslaleh

The ILD Program and objectives are approved by the Queen's University Post-Graduate Medical Office.

The ILD Fellows are involved in the following clinical, academic and research activities:

Clinical activities:

- ILD Clinics and ILD phone consults (at Hotel Dieu Hospital)
- ILD inpatient assessment and follow-up (at Kingston General Hospital)
- Bronchoscopies and BAL
- Transbronchial lung Cryobiopsies.
- Rheumatology Clinics
- Dyspnea Clinics (optional)
- Thoracic Surgery rotation (optional).

A clinical template with a structured approach is used to document clinical encounters in our electronic medical record system (Lumeo).

Academic Activities:

- Monthly ILD Multidisciplinary Rounds
- Monthly ILD Journal club
- Monthly ILD-Rheumatology Rounds
- Weekly BAL results discussions and patient care planning meetings
- Radiology rotation
- Hematopathology Laboratory rotation –to learn how BAL cell differentials are done.
- Optional General Respirology academic activities:
  - Respirology Rounds
  - Respirology Journal Club
  - Respirology Fellows Half Academic days

In addition, our ILD Fellows can do at least one week rotation (as observers) in the ILD clinics either at the University of Toronto under the supervision of Dr. Shane Shapera or at McMaster University under the Supervision of Dr. Nathan Hambly, who have graciously agreed to have our ILD Fellows rotating at their ILD centers.

Academic resources:

- All relevant ILD Guidelines and ILD articles are available to ILD Fellows in a dedicated folder "ILD Fellows Docs" within the KHSC server.
- A research methodology manual is also available in the "ILD Fellows Docs". This manual was created by Dr. Moran and contains the principles of research methods that the ILD Fellows are encouraged to use in their critical appraisal of the articles they present in ILD Journal club, along with other critical appraisal resources/guidelines for specific types of articles (e.g. CONSORT, STARD, STROBE and PRISMA guidelines).

Research activities:

Since its inception, in May 2013, all the data routinely collected in the ILD clinics through standardized questionnaires and protocols on all patients (shown in Appendix 8) have been recorded in a database. This database contains clinical, functional, radiological, laboratorial, and histopathological information, as well as the final diagnosis established in the ILD Clinic, and where applicable in the ILD Multidisciplinary Rounds.

The main data contained in the ILD database are listed below:

Clinical data

- MRC dyspnea questionnaire
- Modified ACCP and "HP-White paper" Questionnaires for Environmental Exposures
- List of drugs associated with ILD
- Connective Tissue Disease Screening Questionnaire
- Physical exam findings: CTD findings, respiratory and cardiac findings, clubbing.

Functional data

- Complete pulmonary function tests: Spirometry, lung volumes and diffusing capacity. At baseline pre- and post-bronchodilator testing is also performed.
- 6-minute walk test (6MWT): Distance walked (absolute and as % of predicted), oxygen saturation at the start and end of the 6MWT and the dyspnea Brog scale.

Radiological data

- Radiological diagnosis of chest CT findings: UIP pattern, probable UIP pattern, Indeterminate UIP pattern, alternative to UIP pattern, or other.
- Radiological abnormalities: Reticulation, honeycombing, traction bronchiectasis, as well as their distribution.

Laboratorial and Histopathological data

- CBC, ESR, CRP, urine microscopy at baseline, and when clinically indicated on follow-up.
- Connective Tissue Disease work up: Antinuclear antibodies (ab), double strand DNA ab, rheumatoid factor, cyclic citrullinated peptide ab, creatine kinase, ANCAs, extractable

nuclear antigen antibodies. Extended myositis panel when clinically indicated or if CK is elevated.

- Bronchoalveolar lavage cell count and differential; flow cytometry and CD4/CD8 ratio if BAL lymphocytosis (>15% lymphocytes) and there is the clinical suspicion of Sarcoidosis or Lymphoma.
- Histopathological diagnosis of transbronchial biopsies / cryobiopsies or surgical lung biopsies.

This information is collected at baseline and/or at each clinic appointment in a standardized fashion and entered routinely into the ILD database by the ILD Fellow or the ILD Nurse.
The information from the ILD database has been used for Dr. Moran and ILD Fellows initiated studies and to estimate the number patients from our ILD clinic we are able to recruit for Pharma initiated studies.

Evaluations and Feedback:

ILD Fellows are evaluated formally by the Director of the ILD Program, Dr. Onofre Moran, and by the ILD Program Faculty when Fellows rotate with them on a monthly or bi-monthly basis through the Queen's University Electronic evaluation system (Elentra). In addition, the required evaluations by the Saudi Arabia or United Arab Emirates Funding Bureaus, etc. are also performed, when applicable.

Feedback is also provided regularly to ILD Fellows by Dr. Moran and the ILD Faculty:

- During ILD clinics, after each case presentation for outpatients.
- During case presentations for patients with ILD admitted to KGH and during follow-up reviews.
- Before bronchoscopies (ILD Fellows present the case and plans are discussed), as well as during and after bronchoscopies.
- Post-bronchoscopy BAL result discussions and patient care planning meetings.
- During the formal review of dictated clinic and bronchoscopy letters on a regular basis during the first trimester and as required afterwards.
- During ILD Multidisciplinary Rounds case presentations and on the dictated ILD notes afterwards.
- During ILD Journal Club and ILD-Rheumatology Rounds (and corresponding dictated notes afterwards).
- During research projects planning and development.

Feedback to the ILD Program through Elentra is obtained from the ILD Fellows to improve the Program.

Vacations and Conferences:

ILD Fellows are entitled to 3 weeks of vacation during the year (maximum 2 weeks at a time).

The ILD Fellows are encouraged to attend 1-2 conferences per year and present their research results. The main expenses to attend conferences are paid by the ILD Program, up to $2,500 CAD, if the ILD fellow is presenting research results at the conference and does not have funding from their sponsor.

ILD Program Collaborators

The following people collaborate with the clinical and/or academic aspects of our ILD Program:

- ILD Specialists: Dr. Onofre Moran and Dr. Kirsten Nesset.
- ILD Fellowship Program Coordinator – Mrs. Sarah Oddell
- ILD Nurse – Pending to define*
- Rheumatology – Dr. Marie Clements-Baker*
- Chest Radiology - Dr. Rob Dhillon* and Dr. Dominique DaBreo*
- Pathology* - Dr. Alexander Boag*
- Thoracic Surgery* -Dr. Kenneth Reid, Dr. Wiley Chung* and Dr. Andrew Giles
- Hematopathology - Dr. David Good
- Palliative Care/Dyspnea Management - Dr. Danielle Kain and other members pf the PC Division
- Pulmonary Rehabilitation - Dr. Alberto Neder
- Research Coordinators/assistants – Pending to define
- University of Toronto - Dr. Shane Shapera
- McMaster University - Dr. Nathan Hambly

In addition, members of the Division of Respiratory and Sleep Medicine collaborate with the training of our ILD Fellows through the Respirology Rounds and Journal Club presentations, as well as the ILD patients hospitalized at KGH under the Respirology Consult Service.

*Annual meetings to review and update our ILD protocols are held with the ILD Program collaborators.

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APPENDICES

Appendix 1

Protocol to apply ROUTINELY to patients referred to ILD clinic

On Initial Visit:

- Modified ACCP / HP environmental exposures questionnaire.
- MRC + dyspnea VAS + King's brief ILD SOB Questionnaires (to mail to patients with appointment or to be filled out by patients before clinic).
- High Resolution chest CT (HRCT) read by Chest radiologist as per ATS/ERS/JRS/ALAT guidelines. For patients with chest CTs done outside KGH/HDH, please have them Upload in PACS and request Consult by Radiologist using the ILD Clinic requisition.
- PFTs routine and post-bronchodilator + 6 min walk test.
- Systematic investigation of symptoms and signs of CTD: Using CTD questionnaire below to investigate Raynaud's, arthralgias or arthritis, pleuritis, alopecia, skin rashes, photosensitivity, myalgias, muscle weakness, dysphagia, etc.
- Routine assessment of medication list and search for their association with ILD in the Pneumotox website. Ask patients to bring list of medications used in past year.
- Routine lab investigations for CTD & Vasculitis:
  ANA, RF, anti CCP-ab, ENA, ANCAs, ESR, CRP, CK, CBC and differential, Cr, urea.
  As required: Anti-GBM ab, ACE levels, extended myositis panel, etc.

  Bronchoscopy for BAL cell differential- if environmental exposures of risk, including feather, foam in bedding or exposure to drugs associated with ILD or exposure to drugs associated with ILD.
- Consider Cryobiopsy/VATS biopsy if Indeterminate or Alternative UIP pattern on CT.

On follow-up visits (every 3-6 months)

- PFTs routine + 6 min walk test.
- CBC and differential, ESR, CRP, **whichever was initially elevated at baseline + during exacerbations.**
- HRCT Chest every 6 months x 2 years; then as per clinical indication.

The questionnaires are mailed to patients before their appointments.
The PFT and HRCT are arranged before the patient's appointment to the ILD clinic. The lab work will be done on / or before the day of the ILD clinic appointment.

Appendix 2

ENVIRONMENTAL AND DRUG EXPOSURES ASSESSMENT

Interstitial Lung Diseases (ILD) Clinic
Exposures Questionnaire

Please fill the following questionnaire as accurate as possible with the best of your knowledge.-

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* Check the single number that describes the point at which you become short of breath:

_____1. I am not troubled with breathlessness except with strenuous exercise

_____2. I get short of breath when hurrying on the level or walking up a slight hill.

_____3. I walk slower than people of my age on the level because of breathlessness or I have to stop from breath when walking on my own pace on the level.
_____4. I stop for breathe after walking about 100 yards (90 meters) (or after a few minutes) on the level.
_____5. I am too breathless to leave the house or breathless on dressing or undressing.

1. **How often do you cough?** (do not include clearing your throat)

_____ Not at all, or only rarely
_____ Occasionally, but not bothersome
_____Most days
_____ Often or in severe attacks that interfere with activity

2. **How long have you been coughing?____**Months/Years/not applicable

3. **Do you cough at night?** Yes_____ No_____

a) If you cough at night, does it awaken you? Yes____ No____

4. **The cough produces: (check all that apply)**

no phlegm_____ phlegm_____blood_____ Don't cough_____

5. **How long ago to you think the shortness of breath began?_____**

6. **Have you ever smoked, inhaled or injected "recreational" drugs? Yes_____ No_____**

(Include "street drugs" or crushed pills. Do not include prescribed inhalers).

Medications

List drugs you regularly take during last year (better if you can provide a printed list from your pharmacy)

DRUG NAME	DOSAGE (mg and times/day)	For how long have you been on this drug (approximate number of months or years)

Medication history

Have you ever taken any of the following medications?

Anti-inflammatory medications:
____ Azathioprine (Imuran)
____ Chlorambucil
____ Colchicine
____ Gold salts
____ Interferon (any
____ Methotrexate
____ Penicillamine
____ Prednisone

Antibiotics/ infection treatment:
____Cephalosporin
____ Isoniazid (INH)
____ Macrolide
____ Minocycline
____ Nitrofurantoin (Macrodantin)
____ Penicillin
____ Sulfonamides (TMP-SMX)

Cancer therapy:
____ Busulfan
____ Bleomycin
____ Cyclophosphamide
____ Etoposide
____ GMCSF
____ Mitomycin
____ Nilutamide
____ Nitrosoureas
____ Radiation
____ Vinblastine

Cardiovascular medications:
____ Amiodarone (Cordarone)
____ Captopril (Capoten)
____ Hydralazine
____ Hydrochlorothiazide
____ Procainamide (Procain SR)
____ Sotolol

Gastrointestinal medications:
____ Azulfidine
____ Sulfasalazine

Miscellaneous medications:
____ Fenfluramine/ dexfenfluramine
____ Leukotriene inhibitor (Singulaire, Accolate)
____ Propylthiouracil
____ Bladder BCG

Neurological medications:
____ Bromocriptine
____ Carbemazepine (Tegretol)
____ L tryptophan
____ Phenytoin (Dilantin)

The ILD Nurse and/or ILD Physicians will ascertain if any of the drugs the patient is on, or has used, have been associated with ILD in Pneumotox.com +/- drug monograph +/- Pubmed.

Exposures in

1. Home, household and/or work place:

Do you/did you have any of the following items/conditions? Tick the box if YES. ***If you answer YES to any question, specify dates (from- till)***

	YES		YES
Foam (sponge) in: **- Pillows** **- Mattress (including in mattress pillow top?** **Please check your pillows and mattress to answer this question.**		**Feather or down in:** **Pillows** **Duvets** **Feather pillows** **Feather blankets** **Feather jackets?** **Bamboo in pillows?**	
Birds **(Include pigeons, doves, parakeets, cockatiels, chickens, ducks, geese, pheasants)**		**Bird droppings** **Clean coups, cages for birds?**	
Visible moulds or mouldy odour in house		**Humidifier or dehumidifier?**	
Flooded house		Furry animals	
Barns, chicken coops or stables?		Hay	
Shoot birds as a hobby and skin or stuff the birds feathers?		Nests in attics	
Wood dust		Change carpets or wooden floors at home?	
Water leaks		Sauna/hot tub/bathtub/jacuzzi	
Indoor swimming pools		Ventilation system at home or work	
Unkept filters		Air cooler or mist fountain	
Unkept old carpets		Air conditioning units	
Unkept heat sources, furnaces		Use hair sprays?	

2. Profession, employment and workplace

A. State all your past and current professions/employments

Profession - employment	**What exactly did you do? Which materials did you work with? State all dusts, fumes and chemicals.**	**From (Year)**	**Till (Year)**

B. Tick the appropriate boxes if you have worked in these industries/professions/workplaces (include dates)

	Yes		**Yes**		**Yes**
Mining industry		Plastic industry		Crop farming/ farmer	
Stone quarry		Carpenter/joiner		Grain mill	
Tunelling		Pulp mill/paper mill		Gardener	
Ceramic industry		Animal/stock husbandry		Dairy/ milkman	
Insulating		Forestry		Brewery/ winery	
Foundry		Woodwork		Chicken/poultry coup	
Saw mills working		Dental technician		Laboratory animals	
Stucco working		Painter		Microbiology lab/ lab Worker	
Car mechanic		Firefighter		Horse stables	
Insulation worker		Plumber/tinner		Hay handling	
Metal working industry		Trash collector/worker		Bakery/ baker	
Turner/miller		Wastes processing		Veterinarian	
Aluminium industry		Detergent production		Mollusc processing	
Glass industry		Wind instruments player		Mushroom growing/picking	
Welder		Barns		Food industry	
Sand blaster		Textile industry		Ironing work	
Nacre processing		Jeweller		Photocopying	

Other exposures in workplace, household, hobbies, neighbourhood

If you have been exposed **repeatedly** to the materials below tick appropriate box and include dates.

Exposure	**Yes**	**Exposure**	**Yes**
Cattle farming		Cheese processing	
Meat processing		Coffee/tee processing	
Vegetable growing and processing		Mushroom processing	
Rapeseed oil		Flour	
Malt		Fish meal	
Oil nasal drops		Cotton	
Cork		Enzymes	
Insecticides		Fertilizers	
Asbestos		Brakes	
Cement		Clay, ceramic	
Silica		Ceramic tiles	
Barium		Beryllium	
Cobalt		Chrome	
Coal		Iron	
Mica		Talc	
Tin		Aluminium	
Isocyanates (foam, sprays, glues)		Colours/dyes	
Metalworking fluids		Industrial cleaning solutions	

List any other exposures that you feel might be related to your lung disease?

_____

_____

Appendix 3

The King's Brief Interstitial Lung Disease Questionnaire (K-BILD)©2011

This questionnaire is designed to assess the impact of your lung disease on various aspects of your life. Please circle the response that best applies to you for each question

1. In the last 2 weeks, I have been breathless climbing stairs or walking up an incline or hill.

1. Every time 2. Most times 3. Several Times 4. Some times 5. Occasionally 6. Rarely 7. Never

2. In the last 2 weeks, because of my lung condition, my chest has felt tight.

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

3. In the last 2 weeks have you worried about the seriousness of your lung complaint?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

4. In the last 2 weeks have you avoided doing things that make you breathless?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

5. In the last 2 weeks have you felt in control of your lung condition?

1. None of the time 2. Hardly any of the time 3. A little of the time 4. Some of the time 5. A good bit of the time 6. Most of the time 7. All of the time

6. In the last 2 weeks, has your lung complaint made you feel fed up or down in the dumps?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

7. In the last 2 weeks, I have felt the urge to breathe, also known as 'air hunger'.

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

8. In the last 2 weeks, my lung condition has made me feel anxious.

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

9. In the last 2 weeks, how often have you experienced 'wheeze' or whistling sounds from your chest?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

10. In the last 2 weeks, how much of the time have you felt your lung disease is getting worse?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

11. In the last 2 weeks has your lung condition interfered with your job or other daily tasks?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

12. In the last 2 weeks have you expected your lung complaint to get worse?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

13. In the last 2 weeks, how much has your lung condition limited you carrying things, for example, groceries?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

14. In the last 2 weeks, has your lung condition made you think more about the end of your life?

1. All of the time 2. Most of the time 3. A good bit of the time 4. Some of the time 5. A little of the time 6. Hardly any of the time 7. None of the time

15. Are you financially worse off because of your lung condition?

1. A significant amount 2. A large amount 3. A considerable amount 4.A reasonable amount 5. A small amount 6. Hardly at all 7. Not at all

Appendix 4

CTD questionnaire - To be answered by <u>New Patients</u> attending the ILD clinic.

Name_____ Date_____

	YES	NO
Have you ever had arthritis or rheumatism for more than 6 weeks?	☐	☐
Do you have morning stiffness in the joints, lasting at least one hour (which joints)?	☐	☐
Do your fingers become white or purple in the cold?	☐	☐
Have you ever had a prominent rash on your cheeks for more than 1 month?	☐	☐
Does your skin break out after you have been in the sun (not sunburn)?	☐	☐
Has it ever been painful to take a deep breath for more than a few days?	☐	☐
Have you ever had rapid loss of lots of hair?	☐	☐
Have you recently had swallowing problems?	☐	☐
Have you recently had muscle weakness or pain like getting in/out of chair/bed?	☐	☐
Have you had recently frequent fevers?	☐	☐
Have you lost weight recently?	☐	☐
Do you have an ongoing sensation of dry/gritty eyes, or do you need to use eye drops daily for dry eyes.	☐	☐
Do you have a dry mouth or need water to help you swallow dry food?	☐	☐

Routine Rheumatology work up.

- Routine lab investigations for CTD & Vasculitis:
  ANA, DsDNA, RF, anti CCP-ab, ENA, ANCAs, ESR, CRP, CK, CBC and differential, Cr, urea.
- Extended Myositis panel: If CK is elevated, clinical findings of Myositis or the patient has Raynaud's or features of antisynthetase syndrome or as indicated by Dr. Clements-Baker after clinical assessment.

Rheumatology Referral Criteria

Refer patients to Rheumatology If:

1. *Clinical suspicion of CTD –based on the ILD questionnaire or physical exam-; or*
2. *ANA ≥1:320 with diffuse, speckled or homogeneous patterns; or ANA with centromere or nuclear pattern ≥1:80.*
3. *Elevated CK with no clear explanation.*
4. *Abnormal ENA antibodies.*

When Myositis is suspected send Mitogen requisition attached to the laboratory

Advanced Diagnostics Laboratory
Mitogen

***This requisition form is ONLY for out of province labs using, please note that Mitogen lab ONLY sends the reports to the referral labs.*

Version: 20180330

Dr. M.J. Fritzler, Director
3330 Hospital Dr. NW: HRIC3A26;
Calgary, AB T2N 4N1
Phone: 403-220-4582 Fax: 403-210-8616
Email: madl@ucalgary.ca
Website: www.mitogen.ca

Autoantibody Test Requisition

Patient Information:	*Referring Physician Information:*
Name: (Surname, First)	*Dr. Name: (Surname, First)*
*PHN :	*Phone:
*DOB: (dd/mm/yy) *Gender:	* Fax# :
*Address:	*Email:
Postal Code:	

Referring Lab Information:	*Sample Information:*
*Lab Name:	*Date/Time collected: (dd/mm/yy: hr)
*Address:	*Diagnostic Information pertinent to autoantibody test request:
*Phone :	Phlebotomy lab: *All tests ordered can be done on serum/CSF from a single SST tube: minimum sample volume 3.0 ml.*
*Fax #	

* = Required information.

Medical Personnel: Please mark **ALL** tests to be done.

*Anti-Cellular Antibodies *(Atypical and Cell Cycle Patterns)* *Anti-dsDNA: quantitative SLE disease activity *Anti-single stranded DNA (ELISA) *Anti-Histone: Drug-Induced Lupus *Anti-DFS (Dense Fine Speckled70/LEDGF)	*Nephritis: -Idiopathic Membranous Nephropathy: Anti-PLA2R (phospholipase A2 receptor) -Primary Membranous Nephropathy: Anti-THSD7A (thrombospondin) *Inflammatory Bowel Disease profile: ASCA (IgG + IgA);	*Encephalitis: - NMDA (NR1) Receptor Antibodies - Anti-DPPX (dipeptidyl aminopeptidase-like 6) - VGKC Antibodies (Voltage gated potassium channel – LGI1 & Caspr2) - Anti-GABAB Receptor - Anti-AMPA Receptor
*Systemic Lupus Profile: anti- Sm , U1RNP, Ro52/TRIM21, SSA/Ro60, SSB/La, PCNA, dsDNA, Chromatin, Ku,Ribo-P.	* Arthritis Panel: -Anti-Citrullinated peptides – anti- HCP1,HCP2,VCP1 and VCP2 -Rheumatoid Factor (IgM) IgA available	*Paraneoplastic Disease Profile: Amphiphysin, Ri (NOVA-1), Yo, Hu, PNMA2 (Ma2/Ta), CV2/CRMP-5, Recoverin, SOX1, Titin. * Idiopathic Ataxia Anti-MPP-1 *(LDT)*
*Scleroderma/Systemic Sclerosis Profile: Anti-CENP A + B, Topo-I/Scl-70, RNA polymerase III, fibrillarin, Th/To, Ku, PDGFR, Ro52/TRIM21, PM/Scl-75, PM/Scl-100, NOR90/hUBF *Sjögren's Syndrome Profile: Anti-SS-A/Ro, SS-B/La, anti-Ro52/TRIM21 *Nucleolar Autoantibody Profile *Nuclear Envelope/Membrane Profile	*Autoimmune Myopathy / Myositis Profile: Jo-1, Mi2, Mi2-α, Mi2β, MDA5, NXP2, TIF1γ PL7, PL12, PM/Scl75, PM/Scl100, Ku, SRP, Ro52, EJ, OJ, Ro52. * Immune Mediated Necrotizing Myopathy and Stain Related Myopathy : -Anti-HMGCR , Anti-SRP *Inclusion Body Myositis: - Anti-NT5C1 A *(LDT)* *Myasthenia Gravis: Anti-AChR	*Neurological Disease Profile (IgG +IgM): Anti-GM1, GM2, GM3, GD1a, GD1b, GT1b, GQ1b *Neuromyelitis Spectrum Profile : (NMO/MOG) Anti-Aquaporin 4/ Anti-Myelin Oligodendrocyte Glycoproteins *Anti-myelin associated glycoproteins (MAG) * Anti-IgLON5 *Anti-GAD 65
*Anti-Phospholipid Syndrome - Anti-Domain 1 β2GPI - Phosphatidylserine/Prothrombin Complex (PS/PT) -IgG, IgM: Lupus Inhibitor/ Anticoagulant Replacement	*Autoimmune Liver Disease Profile: M2/M3, 3EBPO, LKM, SLA, SP100, gp210, PML, LC-1, Ro 52/TRIM21 *Cytoplasmic Dot Profile GW Bodies (Ge-1, GW182, Ago-2); EEA1 *(LDT)* *Cancer Associated Autoantibody Panel: CENP-F, P53 *(LDT)*	*Vasculitis - ANCA/PR3, MPO quantitative by CIA - Atypical ANCA by multiplex: Anti-Lamp2, Anti-Elastase (Levamisole/Cocaine Related) *LDT* - Anti-p140/p155/TRIM28 *(LDT)*
Bullous Autoimmune Skin Disease Profile (BP180, BP230,Desmoglein 1, Desmoglein 3)	*Lung - Interstitial Lung Disease Antibody (ILD) Profile - Alveolar Proteinases :anti-GMCSF (*LDT)*	Other *(specify):*

Please send properly labeled and packaged serum samples with this requisition to:
Mitogen Advanced Diagnostics Laboratory; c/o Dr. MJ Fritzler.
University of Calgary (HRIC 3A26), 3330 Hospital Dr. NW Calgary, AB T2N 4N1

Appendix 5

Chest Imaging Protocols

Table 3. High-Resolution Computed Tomography Scanning Parameters

Recommended Scanning Protocol	Advantages of Updated Recommendations
1. Noncontrast examination	—
2. Volumetric acquisition with selection of: • Sub-millimetric collimation • Shortest rotation time • Highest pitch • Tube potential and tube current appropriate to patient size: ° Typically 120 kVp and ≤240 mAs ° Lower tube potentials (e.g., 100 kVp) with adjustment of tube current encouraged for thin patients • Use of techniques available to avoid unnecessary radiation exposure (e.g., tube current modulation)	A. Acquisition covering the entire lung volume (vs. analysis of 10% of lung volume with sequential scanning) • No risk of missing subtle infiltrative abnormalities • Possibility of multiplanar reformations, helpful for analysis of the ILD pattern and predominant distribution of lung changes • Possibility of post-processing to optimize detection of subtle hypoattenuated lesions (minimum intensity projection) and micronodular infiltration (maximum intensity projection) • Possibility of detection of additional lesions (e.g., incidental identification of lung nodule or focal consolidation in lung fibrosis that may correspond to lung carcinoma) • Optimal to assess progression or improvement in patient's follow-up B. Dramatic increase in temporal resolution and speed of data acquisition • Motion-free images C. Availability of numerous dose-reduction tools
3. Reconstruction of thin-section CT images (≤1.5 mm): • Contiguous or overlapping • Using a high-spatial-frequency algorithm • Iterative reconstruction algorithm if validated on the CT unit (if not, filtered back projection)	—
4. Number of acquisitions: • Supine: inspiratory (volumetric) • Supine: expiratory (can be volumetric or sequential) • Prone: only inspiratory scans (can be sequential or volumetric); optional (*see text*) • Inspiratory scans obtained at full inspiration	A. Expiratory scans useful to detect air trapping B. Prone scans allow analysis of peripheral lung changes without dependent lung atelectasis that may be mistaken for abnormal lung infiltration or mimic disease (e.g., pseudohoneycombing when combined with paraseptal emphysema) C. Inadequate inspiration increases lung attenuation (which should not be interpreted as ground-glass attenuation) and is responsible for dependent lung atelectasis (which may mimic abnormal lung infiltration or mask subtle abnormalities)
5. Recommended radiation dose for the inspiratory volumetric acquisition: • 1–3 mSv (i.e., "reduced" dose) • Strong recommendation to avoid "ultralow-dose CT" (<1 mSv)	A. Considerable dose reduction compared to conventional scanning

Definition of abbreviations: CT = computed tomography; ILD = interstitial lung disease.

Diagnosis of Idiopathic Pulmonary Fibrosis. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline. American Journal of Respiratory and Critical Care Medicine. 198(5): September 1, 2018.

Chest CT protocols for ILD patients

New ILD patients

- **Non contrast examination**
- **Volumetric acquisition 0.5 mm slice thickness**
- **120 kVp, 50 mA with AIDR dose reduction**
- **3 separate acquisitions:**
    - **Supine End Inspiration**
    - **Supine End Expiration**
    - **Prone End Inspiration**
- **Reconstruction 1.0 mm**

1) Axial 1.0 mm Lung Reformat edge enhancement

2) Axial 2.5 mm Lung Reformat

3) Axial, Coronal and Sagittal Mediastinal Reformat

Contiguous slices 2.5 mm.

Repeat ILD patients

- **Non contrast**
- **Volumetric acquisition 0.5 mm slice thickness with dose reduction**
- **Reconstruction 1.0 mm and 2.5 mm Lung**
- **Supine Inspiration**
- **No prone**
- **No expiration**

REQUEST CTS WITHIN 2 WEEKS BEFORE CLINIC APPOINTMENT.

Summary of Chest CT Protocols

ILD New Patient	**ILD Follow up**	**Chest**	**CTPA**
Supine End Inspiration Prone End Inspiration Supine End Expiration	Supine End Inspiration	Supine End Inspiration	Supine End Inspiration
Slice Thickness Lung 1.0 and 2.5 mm axial Mediastinal 2.5 mm axial, coronal and sagittal	Slice Thickness Lung 1.0 and 2.5 mm axial Mediastinal 2.5 mm axial, coronal and sagittal	Slice Thickness Lung 1.0 and 2.5 mm axial Mediastinal 2.5 mm axial, coronal and sagittal	Slice Thickness Lung 1.0 and 2.5 mm axial Mediastinal 1.0 and 2.5 mm axial 2.5 mm coronal and sagittal
Dose DLP 388/5.4 mSV	**Dose DLP 300/4 mSV**	**Dose DLP 300/4 mSV**	**Dose DLP 300/4 mSV**

Interpretation of chest CTs to be done as per Guidelines.

AMERICAN THORACIC SOCIETY DOCUMENTS

Idiopathic Pulmonary Fibrosis (an Update) and Progressive Pulmonary Fibrosis in Adults

An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Luca Richeldi, Carey C. Thomson, Yoshikazu Inoue, Takeshi Johkoh, Michael Kreuter, David A. Lynch, Toby M. Maher, Fernando J. Martinez, Maria Molina-Molina, Jeffrey L. Myers, Andrew G. Nicholson, Christopher J. Ryerson, Mary E. Strek, Lauren K. Troy, Marlies Wijsenbeek, Manoj J. Mammen, Tanzib Hossain, Brittany D. Bissell, Derrick D. Herman, Stephanie M. Hon, Fayez Kheir, Yet H. Khor, Madalina Macrea, Katerina M. Antoniou, Demosthenes Bouros, Ivette Buendia-Roldan, Fabian Caro, Bruno Crestani, Lawrence Ho, Julie Morisset, Amy L. Olson, Anna Podolanczuk, Venerino Poletti, Moisés Selman, Thomas Ewing, Stephen Jones, Shandra L. Knight, Marya Ghazipura, and Kevin C. Wilson; on behalf of the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This official clinical practice guideline was approved by the American Thoracic Society, European Respiratory Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax February 2022

Table 3. High-Resolution Computed Tomography Patterns in Idiopathic Pulmonary Fibrosis

	HRCT Pattern			
	UIP Pattern	**Probable UIP Pattern**	**Indeterminate for UIP**	**CT Findings Suggestive of an Alternative Diagnosis**
Level of confidence for UIP histology	Confident (>90%)	Provisional high confidence (70–89%)	Provisional low confidence (51–69%)	Low to very low confidence (≤50%)
Distribution	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with fibrosis) • Occasionally diffuse • May be asymmetric	• Subpleural and basal predominant • Often heterogeneous (areas of normal lung interspersed with reticulation and traction bronchiectasis/ bronchiolectasis)	• Diffuse distribution without subpleural predominance	• Peribronchovascular predominant with subpleural sparing (consider NSIP) • Perilymphatic distribution (consider sarcoidosis) • Upper or mid lung (consider fibrotic HP, CTD-ILD, and sarcoidosis) • Subpleural sparing (consider NSIP or smoking-related IP)
CT features	• Honeycombing with or without traction bronchiectasis/ bronchiolectasis • Presence of irregular thickening of interlobular septa • Usually superimposed with a reticular pattern, mild GGO • May have pulmonary ossification	• Reticular pattern with traction bronchiectasis/ bronchiolectasis • May have mild GGO • Absence of subpleural sparing	• CT features of lung fibrosis that do not suggest any specific etiology	• Lung findings ◦ Cysts (consider LAM, PLCH, LIP, and DIP) ◦ Mosaic attenuation or three-density sign (consider HP) ◦ Predominant GGO (consider HP, smoking-related disease, drug toxicity, and acute exacerbation of fibrosis) ◦ Profuse centrilobular micronodules (consider HP or smoking-related disease) ◦ Nodules (consider sarcoidosis) ◦ Consolidation (consider organizing pneumonia, etc.) • Mediastinal findings ◦ Pleural plaques (consider asbestosis) ◦ Dilated esophagus (consider CTD)

HP Guidelines to interpret Chest CT

ATS 2020

Chest HRCT Features of Fibrotic HP Pattern

HRCT Pattern	Typical HP	Compatible with HP	Indeterminate for HP
Description	The "typical HP" pattern is suggestive of a diagnosis of HP. It requires *a*) an HRCT pattern of lung fibrosis (as listed below) in one of the distributions and *b*) at least one abnormality that is indicative of small airway disease	"Compatible-with-HP" patterns exist when the HRCT pattern and/or distribution of lung fibrosis varies from that of the typical HP pattern; the variant fibrosis should be accompanied by signs of small airway disease	The "indeterminate-for-HP" pattern exists when the HRCT is neither suggestive nor compatible with a typical and probable HP pattern
Relevant radiological findings	HRCT abnormalities indicative of lung fibrosis are most commonly composed of irregular linear opacities/coarse reticulation with lung distortion; traction bronchiectasis and honeycombing may be present but do not predominate The distribution of fibrosis may be: • Random both axially and craniocaudally or • Mid lung zone–predominant or • Relatively spared in the lower lung zones HRCT abnormalities indicative of small airway disease: • Ill-defined, centrilobular nodules and/or GGOs • Mosaic attenuation, three-density pattern,* and/or air trapping (*often in a lobular distribution*)	Variant patterns of lung fibrosis: • UIP pattern: basal and subpleural distribution of honeycombing with/without traction bronchiectasis (*per 2018 diagnosis of IPF guidelines* [20]) • Extensive GGOs with superimposed subtle features of lung fibrosis Variant (predominant) distributions of lung fibrosis: • Axial: peribronchovascular, subpleural areas • Craniocaudal: upper lung zones HRCT abnormalities indicative of small airway disease: • Ill-defined centrilobular nodules, or • Three-density pattern* and/or air trapping	Lone patterns (i.e., not accompanied by other findings suggestive of HP) of: • UIP pattern (*as per 2018 IPF diagnosis guidelines* [20]) • Probable UIP pattern (*as per 2018 IPF diagnosis guidelines* [20]) • Indeterminate pattern for UIP (*as per 2018 IPF diagnosis guidelines* [20]) • Fibrotic NSIP pattern • Organizing pneumonia–like pattern • Truly indeterminate HRCT pattern

Am J Respir Crit Care Med Vol 202, Iss 3, pp e36–e69, Aug 1, 2020

TABLE 5] Diagnostic CT Categories of Fibrotic HP Based on CT Patterns

HRCT	Typical Fibrotic HP	Compatible With Fibrotic HP	Indeterminate for Fibrotic HP
Features	CT signs of fibrosis with either of the following: • Profuse poorly defined centrilobular nodules of ground-glass opacity affecting all lung zones • Inspiratory mosaic attenuation with three-density sign ***And*** • Lack of features suggesting an alternative diagnosis	CT signs of fibrosis with any of the following: • Patchy or diffuse ground-glass opacity • Patchy, nonprofuse centrilobular nodules of ground-glass attenuation • Mosaic attenuation and lobular air-trapping that do not meet criteria for typical fibrotic HP ***And*** • Lack of features suggesting an alternative diagnosis	CT signs of fibrosis without other features suggestive of HP

CT signs of fibrosis include any of the following: reticular or ground-glass abnormality with traction bronchiectasis; lobar volume loss; honeycombing. The distribution of fibrotic hypersensitivity pneumonitis (HP) is quite variable and often not diagnostically helpful. However, a mid-lung predominant distribution of fibrosis is suggestive of fibrotic HP, and an upper lobe predominance is much more common in fibrotic HP than in idiopathic pulmonary fibrosis. HRCT = high-resolution CT.

Appendix 6

DYSPNEA CLINIC

Referral criteria

1. Dyspnea 4-5/5 on MRC dyspnea scale and
2. Requiring exertional or resting supplementary oxygen; OR
3. Being referred for Pulmonary rehabilitation.

Suggested Initial Management of Dyspnea:

1. Hydromorphone 0.5 mg. q 4 h prn (dyspnea) while awake.
2. Sennokot tabs 8.6 mg. Take 2-4 tablets PO at bedtime or 2 times a day (maximum 8 tabs/day).
3. If required (or if there is abdominal pain), PEG 3350, take 17 gr/day (one heaping tablespoon or one packet) dissolved in 250 ml (1 small glass) of juice, water, soda, coffee or tea; until a BM is achieved.
4. Metoclopramide 5-10 mg PO q 4 h prn (nausea). Use 5 mg for small body built patients.

Avoid Morphine in patients with renal dysfunction and the elderly.

Appendix 7

PULMONARY REHABILITATION REFERRAL CRITERIA FOR ILD CLINIC PATIENTS

All patients at the end are vetted at the rehab clinic and decide whether they are suitable on an individual basis.

The ILD patient who does best is the one with:

- **Clinically stable mild to moderate disease with dyspnea MRC 3/5 (could be MRC 2-4/5; MRC 5 not good candidates*) with reasonable O2 requirements (not exceeding 5lpm during exercise);**
- **Well-motivated;**
- **No significant co-morbid conditions;**
- **No active heart disease**

In summary; the above are general criteria; **every applicant is judged on an individual basis at the Rehabilitation Program in Providence Care**.

- They make exceptions for those on the transplant list and try to expedite admission.
- They don't deny those likely to benefit but try to spare unsuitable patients the grief of failing to respond to exercise training.

Dr. Denis O'Donnell and Dr. Onofre Moran.

Appendix 8

- ILD CLINIC DATA COLLECTION FORM-

PATIENT NAME _____ **CR**_____ **DIAGNOSIS**_____

INITIAL VISIT

	DATE d/m/y
Dyspnea Onset (months/years)	
Dyspnea (MRC)	
Orthopnea/**P**ND/**I**eg edema	
Cough (N/Y) Sputum color	
Cough onset (months/years)	
GERD (N/Y)	
Infectious Sx (N/Y) (**F**ever/**C**hills/**S**weats)	
Constitutional Sx (N/Y) (↓ **W**eight/**A**ppetite/**E**nergy)	
SaO2 on room air (or O2 l/m)	
CTD symptoms (y/n) which? ***as per patient questionnaire***	
Environmental Exposures as per ACCP questionnaire	
Smoker N/Y (pack/yr)	
If ex-smoker, Years Quit	
Environmental exposures Fill adjacent box	
DRUGS associated with ILD See http://www.pneumotox.com/	
Drug 1	
Drug 2	
Drug 3	
PFT results	
FVC (N/%)	/
FEV1 (N/%)	/
FEV1/FVC %	
TLC (N/%)	/
Dlco (N/%)	/
6MWT (m/%)	/
6MWT (SaO_2 start/end)	/
CBC (Hb/WBC)	
ESR (mm/h)	
CRP (mg/l)	

Echo date_____ LVEF_____ PAP_____

other: _____

CHEST CT Date_____ Report: UIP/probable UIP/Indeterminate/Alternative/Other_____

LAB Date_____ RF_____ CCP ab____ ANA_____ DsDNA____ ENA_____ ANCA_____

Bronch Date_____ Path_____ BAL Results: N____ L____ M____ E____

MRC Dyspnea Scale

Grade Description

1 Not troubled by breathlessness except with strenuous exercise.

2 Troubled by shortness of breath when hurrying on the level or walking up a slight hill.

3 Walks slower than people of the same age on the level because of dyspnea or has to stop for breath when walking at own pace on the level.

4 Stops for breath after walking about 100 yards (90 m) or after a few minutes on the level.

5 Too breathless to leave the house or breathless when dressing or undressing.

Environmental Exposures

Y	N	
__	__	Birds
__	__	Feather pillow/duvet
__	__	Foam pillow/duvet
__	__	Mould/Fungus
__	__	Wood work
__	__	Hot tub/Jacuzzi
__	__	Hay / Farm
__	__	Asbestos

Other: _____

FOLLOW-UP VISITS

	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y
Dyspnea **B**etter/**W**orse/**S**ame						
Dyspnea (MRC)						
KB-ILD score						
VAS Dyspnea (cm)						
Orthopnea/**P**ND/**I**eg edema						
Cough (**N**/**Y**) Sputum color						
Cough **B**etter/**W**orse/**S**ame						
GERD (**N**/**Y**)						
Infectious Sx (N/Y) (**F**ever/**C**hills/**S**weats)						
Constitutional Sx (N/Y) (↓**W**eight/**A**ppetite/**E**nergy)						
SaO2 (room air or l/min O2)						
Drugs/exposures w/ILD risk						
MEDICATIONS FOR ILD	**Esbriet__ Ofev____ PPI_____ Other___ _____**	**Esbriet__ Ofev____ PPI_____ Other___ _____**	**Esbriet__ Ofev____ PPI_____ Other___ _____**	**Esbriet__ Ofev____ PPI_____ Other___ _____**	**Esbriet__ Ofev____ PPI_____ Other___ _____**	**Esbriet__ Ofev____ PPI_____ Other___ _____**
FVC (N/%)	/	/	/	/	/	/
TLC (N/%)	/	/	/	/	/	/
Dlco (N/%)	/	/	/	/	/	/
6MWT (m/%)	/	/	/	/	/	/
6MWT (SaO_2 start/end)						
CBC (Hb/WBC)						
ESR (mm/h)						

- ILD CLINIC DATA COLLECTION FORM-

PHYSICAL EXAM

	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y	DATE d/m/y
Crackles	Fine____ Coarse____ None____	Fine____ Coarse____ None____	Fine____ Coarse____ None____	Fine____ Coarse____ None____	Fine____ Coarse____ None____	Fine____ Coarse____ None____
Squeaks	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Wheezes	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Clubbing	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
CHF	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Cor Pulmonale	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
↑ P2	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Arhtritis (specify affected joints)	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Sclerodactyly	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____	Y____ N____
Other findings						
TREATMENT PLAN	**Pirfenidone____Ofev____ PPI_____ Other_______**	**Pirfenidone____Ofev____ PPI_____ Other_______**	**Pirfenidone____Ofev____ PPI_____ Other_______**	**Pirfenidone____Ofev____ PPI_____ Other_______**	**Pirfenidone____Ofev____ PPI_____ Other_______**	**Pirfenidone____Ofev____ PPI_____ Other_______**
	GERD measures____ O2 l/min______ Exposure avoidance ____ Drug avoidance_____ Referrals ____________ **Bronchoscopy_____**	***GERD measures___ O2 l/min______ Exposure avoidance_____ Drug avoidance_____ Referrals ____________***	***GERD measures___ O2 l/min______ Exposure avoidance_____ Drug avoidance_____ Referrals ____________***	***GERD measures___ O2 l/min______ Exposure avoidance_____ Drug avoidance_____ Referrals ____________***	***GERD measures___ O2 l/min______ Exposure avoidance_____ Drug avoidance_____ Referrals ____________***	***GERD measures___ O2 l/min______ Exposure avoidance_____ Drug avoidance_____ Referrals ____________***

Appendix 9

INTERSTITIAL LUNG DISEASES (ILD) CLINIC PATIENT RECOMMENDATIONS

1) **<u>If you have feathers, down, foam or bamboo in your pillows or mattress, change them to anti-allergic types (Polyester or Delcron filled); OR use anti-allergic, anti- dust mite, waterproof mattress and pillowcases.</u>**

2) Avoid exposure to birds, dusts, chemicals or fumes.

3) Remove all visible mold or mildew in walls, ceiling or windows as well as humidifiers and de-humidifiers: Pour vinegar into a spray bottle without watering it down. Spray the vinegar onto the moldy surface and leave it to sit for an hour. Wipe clean the area with water and allow the surface to dry. Alternatively mix 1 part of liquid chlorine bleach with 10 parts of warm water. Sponge or soak stain for 5 to 15 minutes and then rinse.

4) Avoid using bathtubs, hot tubs, jacuzzis and humidifiers, unless your home humidity is low.

5) Keep the indoor humidity between 30% and 50% (check it with a hygrometer).

6) Purchase a pulse oximeter to monitor your blood oxygen levels (oxygen saturation), if you were instructed to do so and **ensure your oxygen saturation is at or above 92% at rest and 88% during exertion**. **Pace yourself or adjust your oxygen as required to achieve this. If your oxygen saturation at rest is 90% or less go to an emergency department.**

7) Follow the anti-acid reflux measures given to you in clinic, **even if you don't have reflux symptoms.**

8) If your ILD Specialist/Respirologist recommended changing some of your medications that could cause lung fibrosis, make sure you arrange an appointment with your family doctor to have those medications changed.

9) If your doctor prescribed a medication for your pulmonary fibrosis, be sure to read the information provided to you about the benefits and potential side effects.

10) Ask your family doctor if your COVID, flu, pneumonia and RSV vaccines are up to date. We recommend that you receive your COVID and flu vaccines annually and the pneumonia vaccines: Pneumovax 23 twice in your lifetime, five years apart, and Prevnar 20 and RSV once in your lifetime.

11) If a procedure (such as a bronchoscopy) is arranged for you make sure that you fully understand the nature of the procedure, the rationale for doing it, and possible complications before you leave clinic.

12) If you are scheduled for a bronchoscopy, make sure that you follow the pre-procedure instructions: such as fasting 6 hours before the procedure and holding blood thinners and some diabetes medications. Use all other medications as usual.

13) Make sure that you fill out the questionnaires that will be sent to you with your next appointment and bring them with you to the clinic appointment.

14) If you feel that your condition is worsening (either your shortness of breath or oxygen saturation), please call Dr. Moran's office at (613) 548-1380 OR **go to an emergency department if your oxygen saturation is 90% or lower at rest.**

Appendix 10

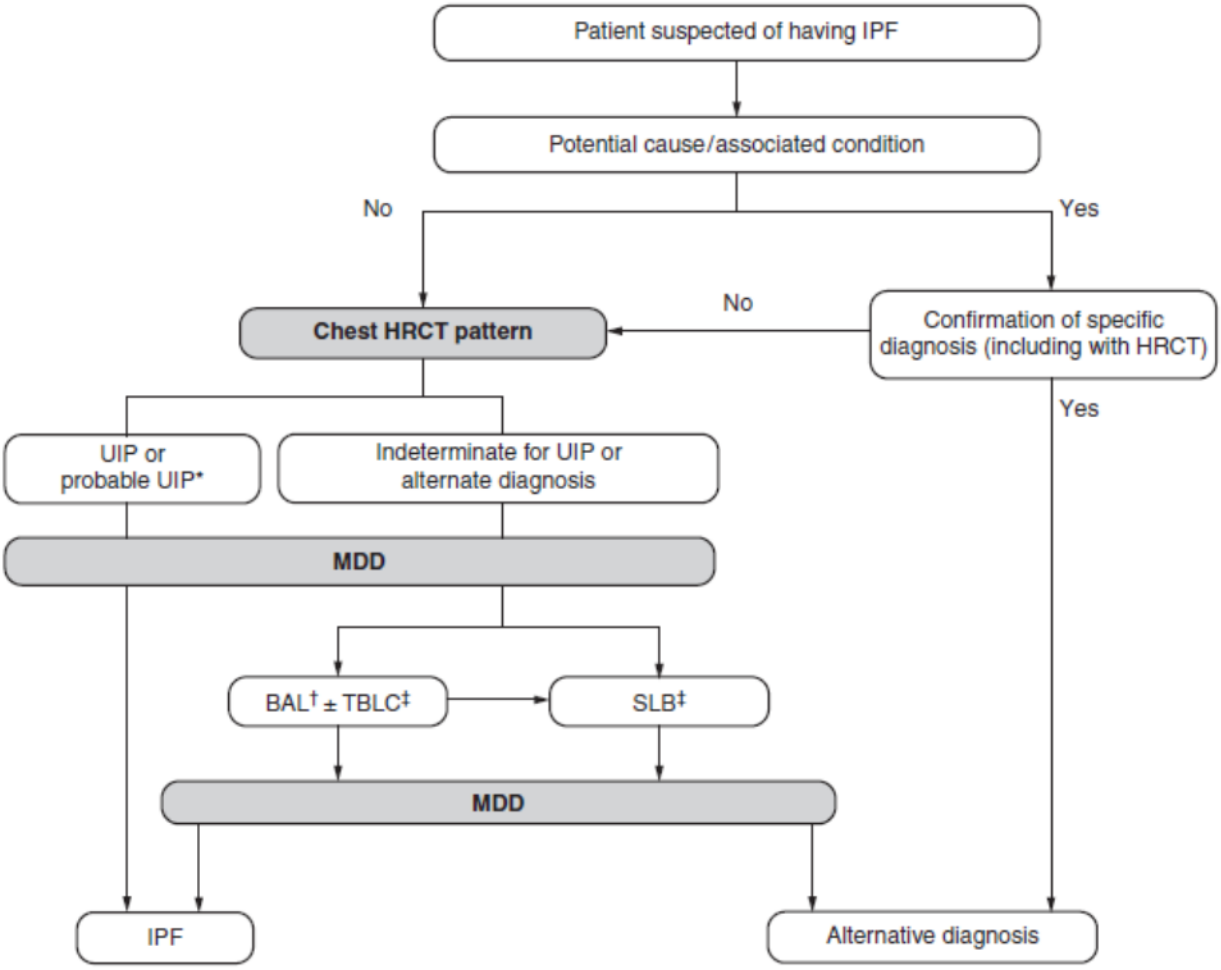
ILD MULTIDISCIPLINARY ROUNDS

Cases to discuss in ILD Multidisciplinary Rounds

Patients with lung biopsy (regular transbronchial biopsies, Cryobiopsies and surgical lung biopsies), are discussed in ILD Rounds by Pathologist, Chest Radiologists and Respirologists applying current guidelines.

All other cases are discussed directly with Chest Radiologists when appropriate: e.g. discrepancy in the diagnosis of UIP patterns on Chest CT between Respirologist and General or Chest Radiologist.

Diagnostic algorithm for IPF



When to perform lung biopsy in patients with suspected HP

[Diffuse Lung Disease Guideline and Consensus Statement]

CHEST

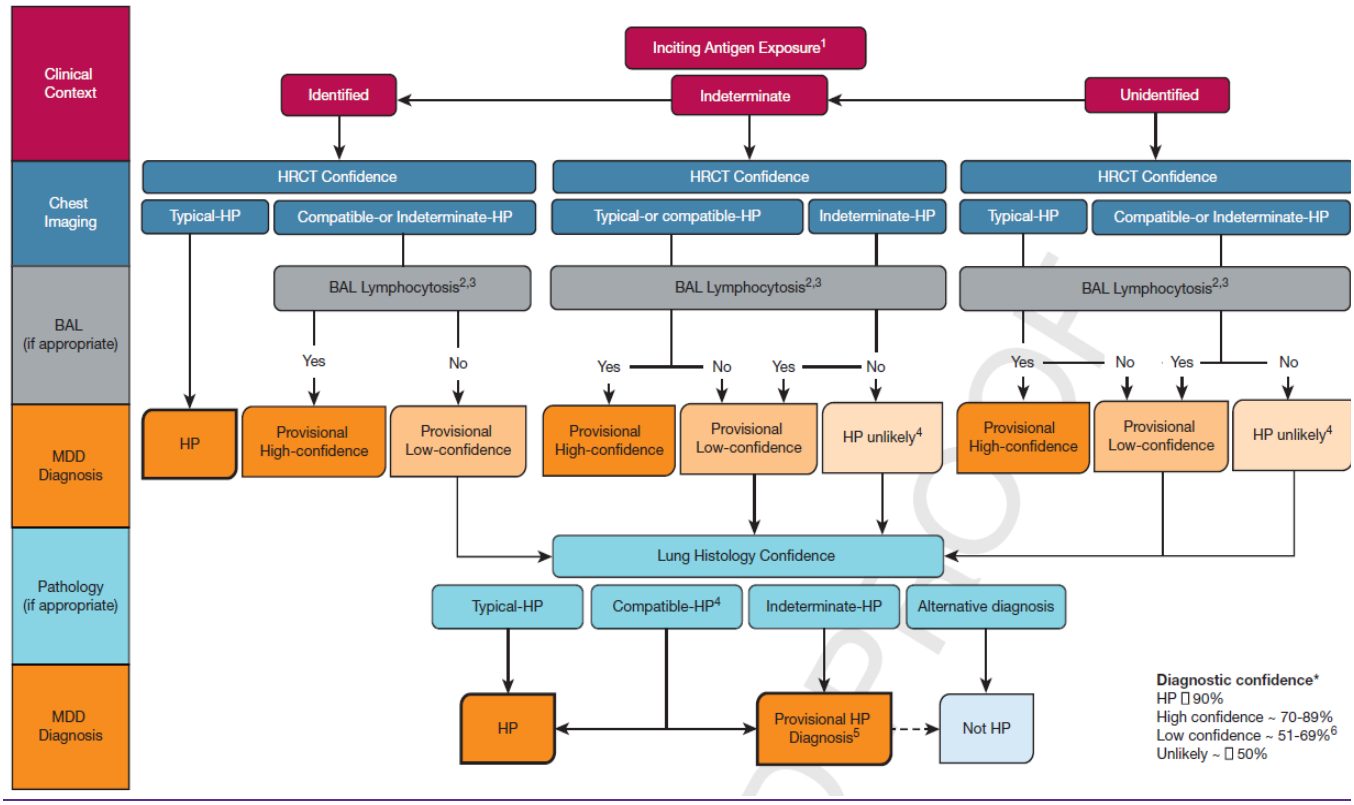
Executive Summary Diagnosis and Evaluation of Hypersensitivity Pneumonitis

CHEST Guideline and Expert Panel Report

Evans R. Fernández Pérez, MD, FCCP; William D. Travis, MD, FCCP; David A. Lynch, MB, BCh; Kevin K. Brown, MD, FCCP; Kerri A. Johannson, MD, MPH; Moisés Selman, MD; Jay H. Ryu, MD, FCCP; Athol U. Wells, MD, FRCP; Yuh-Chin Tony Huang, MD, MHS, FCCP; Carlos A. C. Pereira, MD, FCCP; Mary-Beth Scholand, MD, FCCP; Ana Villar, MD, PhD; Naohiko Inase, MD, PhD; Richard B. Evans, MD, MPH, FCCP;

CHEST 2021; 160(2): e97-e156

Algorithm for the diagnosis of fibrotic and non-fibrotic HP


Clinical Context
Chest Imaging
BAL (if appropriate)
MDD Diagnosis
Pathology (if appropriate)
MDD Diagnosis
Inciting Antigen Exposure1
Identified
Indeterminate
Unidentified
HRCT Confidence
Typical-HP
Compatible-or Indeterminate-HP
Typical-or compatible-HP
Indeterminate-HP
BAL Lymphocytosis2,3
Yes
No
HP
Provisional High-confidence
Provisional Low-confidence
HP unlikely4
Lung Histology Confidence
Compatible-HP4
Alternative diagnosis
Provisional HP Diagnosis5
Not HP
Diagnostic confidence*
HP ~ 90%
High confidence ~ 70-89%
Low confidence ~ 51-69%6
Unlikely ~ 50%

AMERICAN THORACIC SOCIETY DOCUMENTS

Diagnosis of Hypersensitivity Pneumonitis in Adults

An Official ATS/JRS/ALAT Clinical Practice Guideline

Ganesh Raghu, Martine Remy-Jardin, Christopher J. Ryerson, Jeffrey L. Myers, Michael Kreuter, Martina Vasakova, Elena Bargagli, Jonathan H. Chung, Bridget F. Collins, Elisabeth Bendstrup, Hassan A. Chami, Abigail T. Chua, Tamera J. Corte, Jean-Charles Dalphin†, Sonye K. Danoff, Javier Diaz-Mendoza, Abhijit Duggal, Ryoko Egashira, Thomas Ewing, Mridu Gulati, Yoshikazu Inoue, Alex R. Jenkins, Kerri A. Johannson, Takeshi Johkoh, Maximiliano Tamae-Kakazu, Masanori Kitaichi, Shandra L. Knight, Dirk Koschel, David J. Lederer, Yolanda Mageto, Lisa A. Maier, Carlos Matiz, Ferran Morell, Andrew G. Nicholson, Setu Patolia, Carlos A. Pereira, Elisabetta A. Renzoni, Margaret L. Salisbury, Moises Selman, Simon L. F. Walsh, Wim A. Wuyts, and Kevin C. Wilson; on behalf of the American Thoracic Society, Japanese Respiratory Society, and Asociación Latinoamericana de Tórax

This guideline is dedicated to the memory of Prof. Jean-Charles Dalphin† (June 2, 1956–October 17, 2019)

THIS OFFICIAL CLINICAL PRACTICE GUIDELINE WAS APPROVED BY THE AMERICAN THORACIC SOCIETY, JAPANESE RESPIRATORY SOCIETY, AND ASOCIACIÓN LATINOAMERICANA DE TÓRAX MAY 2020

Am J Respir Crit Care Med 2020;202 (3): e36–e69.

HRCT

	Typical for HP		Compatible with HP		Indeterminate for HP	
History of exposure and/or serum IgG testing	Exposure +	Exposure –	Exposure +	Exposure –	Exposure +	Exposure –
No BAL or BAL without lymphocytosis and either no histopathology or indeterminate histopathology	Moderate confidence	Low confidence	Low confidence	Not excluded	Not excluded	Not Excluded
BAL lymphocytosis without histopathology sampling	High confidence	Moderate confidence	Moderate confidence	Low confidence	Low confidence	Not excluded
BAL lymphocytosis with indeterminate histopathology	Definite	High confidence	Moderate confidence	Moderate confidence	Low confidence	Not excluded
Probable HP histopathology	Definite	High confidence	High confidence	Moderate confidence	Moderate confidence	Low confidence
Typical HP histopathology	Definite	Definite	Definite	Definite	Definite	High confidence*

THORACIC SURGERY

Indications for referral - defined with Thoracic Surgery (Dr. Andrew Giles) in our January 2025 annual ILD meeting.

- Non diagnostic Cryobiopsy.
- Cryobiopsy not desired by the patient.
- Need for Dx of a concomitant procedure.
- High risk patients for Cryobiopsy: Morbid obesity, severe PHN, severe/diffuse emphysema, hypercapnic or hypoxic respiratory failure.

Algorithm for IPF diagnosis on the basis of HRCT and biopsy patterns

IPF suspected*		Histopathology pattern†			
		UIP	Probable UIP	Indeterminate for UIP or biopsy not performed	Alternative diagnosis
HRCT pattern	UIP	IPF	IPF	IPF	Non-IPF dx
	Probable UIP	IPF	IPF	IPF (Likely)‡	Non-IPF dx
	Indeterminate	IPF	IPF (Likely)‡	Indeterminate§	Non-IPF dx
	Alternative diagnosis	IPF (Likely)‡	Indeterminate§	Non-IPF dx	Non-IPF dx

Am J Respir Crit Care Med Vol 205, Iss 9, pp e18–e47, May 1, 2022

PATHOLOGICAL CRITERIA FOR DIAGNOSING UIP IN TRANSBRONCHIAL LUNG CRYOBIOPSY

Cryobiopsy for Identification of Usual Interstitial Pneumonia and Other Interstitial Lung Disease Features: Further Lessons from COLDICE, a Prospective Multi-Center Study

Wendy A. Cooper[12,13], Annabelle Mahar[1], Jeffrey L. Myers[4], Christopher Grainge[5], Tamera J. Corte[26,27], Jonathan P. Williamson[28], Michael P. Vallely[2], Simon Lai[10], Ellie Mulyadi[14], Paul J. Torzillo[29], Martin J. Phillips[9,21], Edmund M.T. Lau[28], Ganesh Raghu[17]*, Lauren K. Troy[26]*.

Am J. Respir Crit Care Med. 2020 Dec 7. doi: 10.1164/rccm.202009368OC. Online ahead of print.

Results

- Patchy fibrosis
- Fibroblastic foci" and
- Absence of absence of features to suggest an alternative diagnosis"

in TBC were strongly predictive of UIP at SLB (OR 23.4; p<0.0001).

Current practice at KHSC: Obtain at least 4 biopsies from 2 different segments in the same lobe and consider samples from two different lobes, as surgeons do.