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 now includes:

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

ILD Clinic

The ILD clinics are currently held at Hotel Dieu Hospital from 8:30 am to 1 pm on Thursdays and from 1 pm to 6 pm on Mondays. 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 to 2 ILD clinics and Programs in Munich, Germany.

All diagnostic and management protocols and procedures were 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 ILDs[8].

Following current ILD/IPF Guidelines and with collaboration and input from our colleagues from Chest Radiology, Thoracic Surgery, Rheumatology, 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 the following standardized assessments to properly investigate the most common groups of ILDs: Hypersensitivity Pneumonitis, Connective Tissue Diseases (CTD), and drugs, as per Guidelines[1]:

- Questionnaires:
  -American College of Chest Physicians (ACCP) ILD environmental exposures Questionnaire combined with the "white paper" on Hypersensitivity pneumonitis (shown in Appendix 2)[9,10]
  -King's Brief ILD Questionnaire -pending copyright permission- (see Appendix 3).
  -Connective Tissue Diseases Screening Questionnaire (see Appendix 4).

- High Resolution Chest CT (HRCT) with routine inspiratory, expiratory and prone views (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 their ILD is related to a CTD.

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 diagnostic bronchoalveolar lavage (BAL) procedure, transportation, sample processing, and analysis are performed as per the BAL in ILD Guidelines[8]. The BAL cell count and differential, essential in the diagnostic approach to ILDs, 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.

Once the diagnosis is established, if anti-fibrotic treatment is required, patients are re-scheduled in clinic 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 Physician 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 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 Appendix 7.
-Lung Transplant Program in Toronto: eligible patients diagnosed with IPF or with another advanced or progressive ILD 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 of the ILD patients' HRCTs-, full pulmonary function tests, 6 minute walk tests, and blood work if 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.

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

- Chest Radiology: Dr. Rob Dhillon, Dr. Marina Pourafkari, and Dr. Dominique D'Abreo, 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. All other Respirologists in the Division of Respirology at Queen's University are also invited to attend.
- ILD Nurse: Helps coordinating the follow-up and management plans of patients after rounds.
- ILD Fellows, Respirology Fellows and House Staff rotating in Respirology attend these rounds and present the cases following a standardized Power Point template.
- Thoracic Surgery: Dr. Kenneth Reid, Dr. Willey Chang 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 180 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 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, ensures these plans are implemented. To enhance participation of the house staff in the ILD rounds, lunch is routinely provided.
The diagnostic approach used in our ILD multidisciplinary rounds is shown in Appendix 10.

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

The Queen's University approved ILD Program definition and objectives are provided in a separate document.

The ILD Fellows at Queen's University are involved in the following clinical and academic activities:

Clinical activities:

- ILD Clinics
- ILD inpatient assessment and follow-up
- Bronchoscopies performed on our ILD patients –with performance of BAL and transbronchial cryobiopsies.
- Rheumatology Clinics
- Dyspnea Clinics
- Thoracic Surgery rotation

Academic Activities:

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

In addition, our ILD Fellows can do at least one week rotation 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.

Evaluations and Feedback:

ILD Fellows are evaluated formally by the Director of the ILD Program, Dr. Onofre Moran, on a monthly basis through the Queen's University Electronic evaluation system. Required evaluations are also provided to the Saudi Arabia or United Arab Emirates Funding Bureaus, etc. when applicable.

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

- 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.
- During ILD Journal Club and ILD-Rheumatology Rounds.
- During research projects planning and development.

ILD Research Program

Since its inception, in May, 2013, all the data routinely collected in the ILD clinic through standardized questionnaires and protocols on all patients (Shown in Appendices 1-5) 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/"HP-White paper" Questionnaire for Environmental Exposures of Risk
- Connective Tissue Disease Screening Questionnaire
- List of drugs associated with ILD
- 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.

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, plus on follow-up if clinically indicated.
- 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.
- Bronchoalveolar lavage cell count and differential; flow cytometry and CD4/CD8 ratio if BAL lymphocytosis (>15% lymphocytes).
- 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.
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.

ILD Program Collaborators

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

- ILD Program Coordinator – Mrs. Sarah Oddell
- ILD Nurse - Mrs. Carla Paredes*
- Chest Radiology - Dr. Rob Dhillon*, Dr. Marina Pourafkari* and Dr. Dominique D'Abreo*
- Pathology* - Dr. Alexander Boag*
- Thoracic Surgery* -Dr. Kenneth Reid, Dr. Wiley Chung* and Dr. Andrew Giles
- Hematopathology - Dr. David Good
- Rheumatology – Dr. Marie Clements-Baker*
- Palliative Care/Dyspnea Management - Dr. Danielle Kain and other members pf the PC Division
- Pulmonary Rehabilitation - Dr. Alberto Neder
- Research Coordinators/assistants – Dr. Elias Ordaz and Mrs. Carla Paredes
- 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.

*Meetings to review and update our ILD protocols are held annually with these ILD Program collaborators.

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APPENDICES

Appendix 1

Protocol to be applied 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 (mailed 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 Chest CT 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.
  Write in back of lab req "ANA by IFA form must accompany sample"*As required: Extended Myositis panel, Anti-GBM ab, ACE levels, etc.*

Bronchoscopy for BAL cell count and differential- if environmental exposures of risk, including feather, down or foam in bedding or exposure to drugs associated with ILD.

Consider Cryobiopsy / Referral to Thoracic Surgery 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 elevated at baseline (Our Secretary will check this) + 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 (if none available in last 6 months) 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.

Rheumatology work up

- Routine lab investigations for CTD & Vasculitis:
  ANA **ordered in the initial requisition by IFA** ***Write in back of lab req "ANA by IFA form must accompany sample".*** DsDNA, RF, anti CCP-ab, ENA, ANCAs, ESR, CRP, CK, CBC and differential, Cr, urea, **stop ordering urine analysis**.
- Extended Myositis panel: If CK is elevated, clinical findings of Myositis or the patient has Raynaud's or features / 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 2

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

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

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

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

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

Gastrointestinal medications:
____ Azulfidine
____ Sulfasalazine

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

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/pale, purple, numb, or uncomfortable 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 had recently 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?	☐	☐

Appendix 5

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**
- **Dose > 1000 mGy-cm or 8-10 mSv**
- **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**
- **Dose 300 - 500 mGy-cm or 2-4 mSv**
- **Supine Inspiration**
- **No prone**
- **No expiration**

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 1000/ 14mSV	**Dose DLP 300/4 mSV**	**Dose DLP 300/4 mSV**	**Dose DLP 300 or 4 mSV**

Interpretation of chest CTs will 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)

Appendix 6

DYSPNEA CLINIC

Meeting March, 2018

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.

Symptoms Questionnaire used: Edmonton System Assessment System (ESAS)

1.Watanabe SM, Nekolaichuk C, Beaumont C, Johnson L, Myers J, Strasser F. A multi-centre comparison of two numerical versions of the Edmonton Symptom Assessment System in palliative care patients J Pain Symptom Manage 2011; 41:456-468.
2. Bruera E, Kuehn N, Miller MJ, Selmser P, Macmillan K. The Edmonton Symptom Assessment System (ESAS): a simple method for the assessment of palliative care patients. J Palliat Care 1991; 7:6-9.

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.

Appendix 7

PULMONARY REHABILITATION REFERRAL CRITERIA FOR ILD CLINIC PATIENTS*

Modified as per ILD meeting in May, 2016

We vet all patients 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 (could be MRC 2 to 4; 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**

We make exceptions for those on the transplant list and try to expedite admission; everyone is assessed by both the physios and myself....sometimes ILD patients are admitted briefly purely on compassionate grounds

In summary; the above are general criteria; Dr. O'Donnell **prefered not to provide "hard" entry criteria because this can be misleading and every applicant is judged on an individual basis at the clinic**...we 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.

* Original Meeting- September, 2015.

Appendix 8

- ILD CLINIC DATA COLLECTION FORM-

PATIENT NAME _____ CR_____ **DIAGNOSIS_____**

INITIAL VISIT

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____

	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)	

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)						
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)						
CRP (mg/l)						

- 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 flu and pneumonia vaccines are up to date. We recommend that you receive your flu vaccine annually and the pneumonia vaccines: Pneumovax 23 twice in your lifetime, five years apart and Prevnar 13 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 less at rest.**

Appendix 10

Multidisciplinary case discussions in ILD Rounds

We present in ILD Rounds ONLY cases with surgical lung biopsies, as all other cases can be discussed directly with chest radiologists.

Patients with Surgical lung biopsy +/- bronchoscopy are discussed in ILD Rounds by Pathologist, Chest Radiologists, Respirologist applying current guidelines.

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

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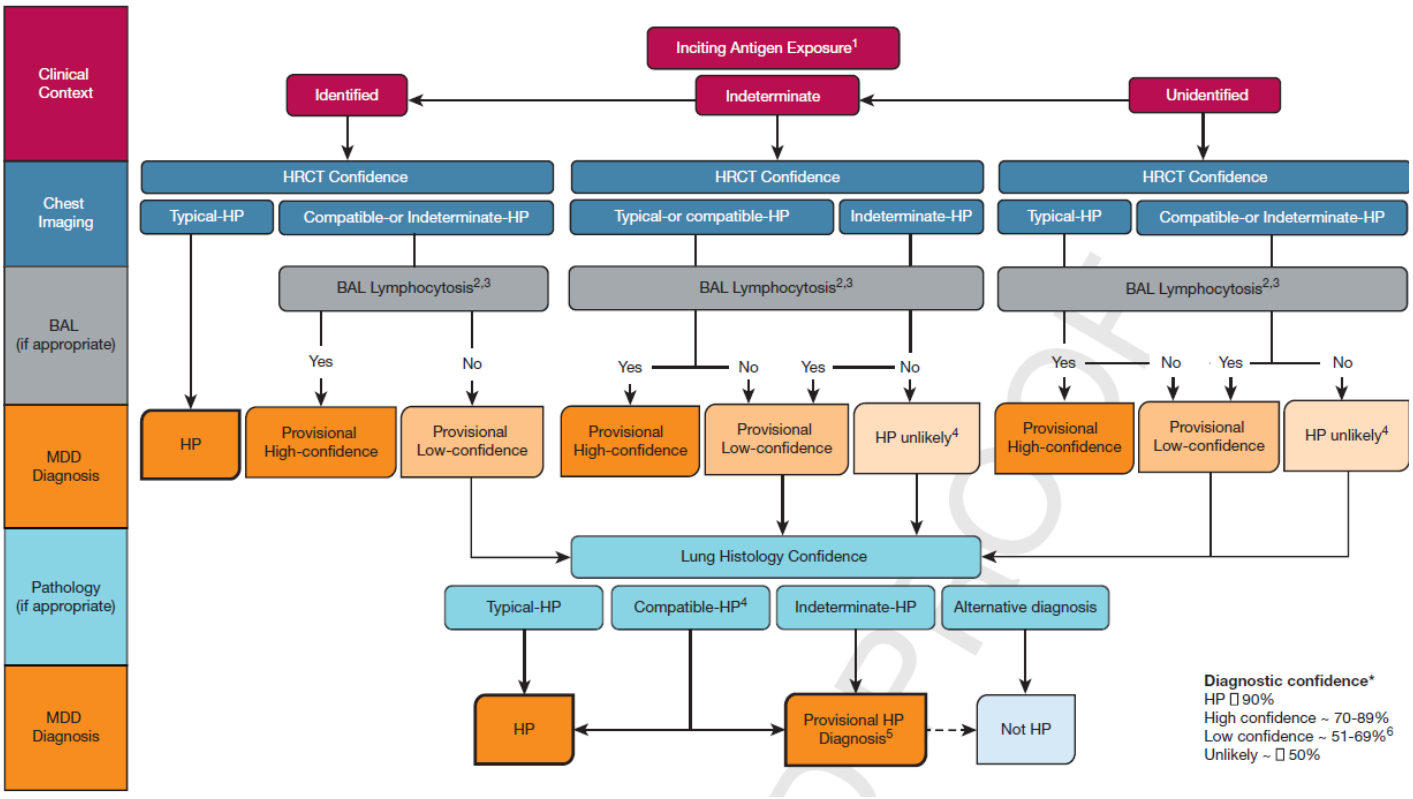
Algorithm for the diagnosis of fibrotic and non-fibrotic HP



Diagnosis of HP in Adults. Am J Respir Crit Care Med Vol 202, Iss 3, pp e36–e69, Aug 1, 2020