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 2 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.

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)[1,2] as well as International Guidelines for the Diagnosis of Idiopathic Interstitial Lung Diseases[3] and the guidelines on Bronchoalveolar Lavage (BAL) in ILDs[4].

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 (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 Questionnaire (shown in Appendix 2)
- Dyspnea Questionnaires: MRC dyspnea questionnaire and University of California at San Diego (UCSD) Dyspnea Questionnaire (see Appendix 3).
-Quality of Life (QOL) questionnaire: St. George's QOL questionnaire.
-Connective Tissue Diseases Screening Questionnaire (see Appendix 4).

- High Resolution Chest CT (HRCT) with routine inspiratory and expiratory 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.

The diagnostic bronchoalveolar lavage (BAL) procedure, transportation, sample processing, and analysis are performed as per the BAL in ILD Guidelines[4]. 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 the 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 Dr. Ingrid Harle, Palliative Care Physician who has kindly agreed to collaborate with our ILD clinic and to manage the dyspnea in our patients. Referral criteria have been established in common agreement (shown in Appendix 6).
- Pulmonary Rehabilitation Program, with Dr. Denis O'Donnell, to assess suitability for enrollment in the rehabilitation program he supervises at Providence Care currently (previously at St. Mary's of the Lake). Referral criteria to this program 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 or their progression-, 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.

ILD Nurse

An ILD Nurse –Mrs. Lynda McCarthy- was hired in January, 2015 to work at the ILD Clinic

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 all 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 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 (performed 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.). She also ensures patients get the proper testing before bronchoscopy (coagulation tests and electrocardiogram) when applicable.

Mrs. McCarthy routinely instructs our ILD patients in clinic about the proper use of a personal pulse oximeter and the use of a peak flow meter, if these are recommended to the patient in the ILD clinic.

Mrs. McCarthy 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 Guidelines[1]-.
The following disciplines collaborate with patients presented in the ILD rounds:

- Thoracic Surgery: Dr. Kenneth Reid and Dr. Willey Chang, who perform the video assisted surgical lung biopsies, as per the "Guidelines" recommendations[1].
- Chest Radiology: Dr. Rob Dhillon, Dr. Justin Flood, and Dr. Dominique D'Abreo, who read all the HRCTs and discuss the radiological findings in 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: Mrs. Lynda McCarthy. Hel;ps coordinating the follow-up and management plans of patients after rounds.
- ILD Fellows, Respirology Fellows and House Staff rotating in Respirology attend these rounds.

The ILD rounds occur monthly and Dr. Moran creates the rounds schedule biannually. To date, more than 70 ILD cases have been presented and discussed in the ILD Multidisciplinary Rounds.

Dr. Moran and/or 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 who is blinded to previous interpretations of the HRCTs. Then, Dr. Boag discusses the histopathological findings from the VATS biopsy. Finally, based on the diagnostic classifications recommended by the Guidelines[1,3], a final diagnosis is achieved in common agreement. Dr. Moran dictates a note that will appear as an addendum in the patient's chart with each of the following diagnoses: Radiological, Pathological and Multidisciplinary diagnosis; with the corresponding follow-up and management plans, as per the Guidelines[2]. Our ILD nurse, who attends the ILD rounds, ensures these plans take place.
To enhance participation of the house staff in the ILD rounds, lunch is provided; this has been secured by Dr. Moran from the two pharmaceutical companies (Boerhinger Ingelheim and Roche) that make the only antifibrotics currently available.

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 been admitted in our program:

-2014-15 Dr. Muhannad Hawari.
-2016-17 Dr. Sami Alyami
-2017-18 Dr. Sharina Aldhaheri (ILD Fellow) and Dr. Sami Alyami (ILD Research Fellow).

The Queen's University approved ILD Program definition and objectives are shown in Appendix 10.

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 biopsies.
- Rheumatology Clinics
- Advanced Dyspnea Clinics
- Thoracic Surgery rotation

Academic Activities:

- ILD Multidisciplinary Rounds
- Weekly post-Bronchoscopy patient discussions and care planning meetings
- Monthly ILD Journal club
- Respirology Rounds
- Respirology Journal Club
- Respirology Fellows Half Academic days
- Radiology Rounds for Respirology and ILD Fellows
- 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 either at the University of Toronto under the supervision of Dr. Shane Shapera or at McMaster University under the Supervision of Dr. Nathan Humbley, who have graciously agreed to have our ILD Fellows rotate 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 Medtech evaluation system. Required evaluations are also provided to the Saudi Arabia or United Arab Emirates Funding Bureaus, 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 patient discussions and care planning meetings.
- After the 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, Respirology Rounds and Journal Club.
- 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 Multidiscilplinary Rounds.

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

Clinical data

- MRC dyspnea questionnaire
- University of California Shortness of Breath Questionnaire
- St. George's Questionnaire
- ACCP 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, possible UIP pattern, Inconsistent UIP pattern, other.
- Radiological abnormalities: Reticulation, honeycombing, traction bronchiectasis, as well as their distribution.

Laboratorial and Histopathological data

- CBC, ESR, CRP, urine microscopy.
- 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 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 Nurse - Mrs. Lynda McCarthy*
- Chest Radiology - Dr. Rob Dhillon*, Dr. Justin Flood* and Dr. Dominique D'Abreo*
- Histopathology* - Dr. Alexander Boag*
- Thoracic Surgery* -Dr. Kenneth Reid and Dr. Wiley Chung*
- Hematopathology - Dr. David Good
- Rheumatology – Dr. Marie Clements-Baker*
- Palliative Care/Dyspnea Management - Dr. Ingrid Harle
- Pulmonary Rehabilitation - Dr. Denis O'Donnell
- Research Coordinators/assistants - Lynda Mc Carthy and Gabriel Abad
- University of Toronto - Dr. Shane Shapera
- McMaster University - Dr. Nathan Humbley

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 set up the ILD protocols were held with the collaborators above at the start of the program (unless specified otherwise in the corresponding Appendices) and in May, 2017.

References

1. Raghu G, Collard HR, Egan JJ, et al. An official ATS/ERS/JRS/ALAT statement: idiopathic pulmonary fibrosis: evidence-based guidelines for diagnosis and management. Am J Respir Crit Care Med 2011;183:788-824.
2. Raghu G, Rochwerg B, Zhang Y, et al. An Official ATS/ERS/JRS/ALAT Clinical Practice Guideline: Treatment of Idiopathic Pulmonary Fibrosis. An Update of the 2011 Clinical Practice Guideline. Am J Respir Crit Care Med 2015;192:e3-19.
3. Travis WD, Costabel U, Hansell DM, et al. An official American Thoracic Society/European Respiratory Society statement: Update of the international multidisciplinary classification of the idiopathic interstitial pneumonias. Am J Respir Crit Care Med 2013;188:733-48.
4. Meyer KC, Raghu G, Baughman RP, et al. An official American Thoracic Society clinical practice guideline: the clinical utility of bronchoalveolar lavage cellular analysis in interstitial lung disease. Am J Respir Crit Care Med 2012;185:1004-14.

APPENDICES

Appendix 1

Approach to be applied ROUTINELY to patients referred to ILD clinic.

On Initial Visit:

- ACCP + St. George's Respiratory Questionnaire + UCSD 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, seizures, fevers or weight loss.
- 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, urinalysis.

  As required: precipitins against bird serum or droppings, Anti-GBM ab, ACE levels...

Referrals to Rheumatology: If clinical suspicion of CTD and/or ANA 1:320 or higher.

Bronchoscopy for BAL cell count and differential-as per protocol that adheres to guidelines if environmental exposures of risk, including feather, down or foam in bedding.

Referral to Thoracic Surgery if NOT typical UIP pattern on CT and IPF is in the differential.

The questionnaires will be mailed to patients before their appointments. The PFT and HRCT (if none available in last 6 months) will be arranged before the patient's appointment to the ILD clinic. The lab work will be done the day of the ILD clinic appointment.

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

- St. George's Respiratory Questionnaire + UCSD Shortness of Breath Questionnaire (to mail to patients with appointment).
- PFTs routine + 6 min walk test.
- CBC and differential, ESR, CRP.
- HRCT Chest every 6 months x 2 years; then as per clinical indication.

The questionnaires will be mailed to patients before their appointments. The PFT and HRCT will be arranged for the patient in the follow-up ILD clinic appointment.
The lab work will be done the day of the ILD clinic appointment.

Rheumatology Referral Criteria

Referrals to Rheumatology: If clinical suspicion of CTD and/or ANA 1:320 or higher.

ANA with centromere nucleolar pattern 1:80

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.

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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
Birds (Include pigeons, doves, parakeets, cockatiels, chickens, ducks, geese, pheasants)		Bird droppings Clean coups, cages for birds?	
Feather duvets, pillows, blankets, jackets ?		Foam in pillows, mattress (including foam top in mattress), elsewhere	
Furry animals		Humidifier	
Flooded house		Visible moulds or mouldy odour in house	
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 in case you have worked in below stated 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 worker	
Car mechanic		Firefighter		Horse stables	
Insulation worker		Plumber/tinner		Hay handling	
Metal industry working		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 (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

UCSD MEDICAL CENTER
PULMONARY REHABILITATION PROGRAM
SHORTNESS-OF-BREATH QUESTIONNAIRE

Please rate the breathlessness you experience when you do, or if you were to do, each of the following tasks. **Do not skip any items**. If you've never performed a task or no longer perform it, give your best estimate of the breathlessness you would experience while doing that activity. Please review the two sample questions below before turning the page to begin the questionnaire.

When I do, or if I were to do, the following tasks, I would rate my breathlessness as:

0	None at all
1	
2	
3	
4	Severe
5	Maximal or unable to do because of breathlessness

1. Brushing teeth ..... **0 1 2 ③ 4 5**

Harry has felt moderately short of breath during the past week while brushing his teeth and so circles a three for this activity.

2. Mowing the lawn ..... **0 1 2 3 4 ⑤**

Anne has never mowed the lawn before but estimates that she would have been too breathless to do this activity during the past week. She circles a five for this activity.

UCSD Shortness of Breath Questionnaire

When I do, or if I were to do, the following tasks, I would rate my breathlessness as:

0	None at all
1	
2	
3	
4	Severe
5	Maximal or unable to do because of breathlessness

1.	At rest	0	1	2	3	4	5
2.	Walking on a level at your own pace	0	1	2	3	4	5
3.	Walking on a level with others your age	0	1	2	3	4	5
4.	Walking up a hill	0	1	2	3	4	5
5	Walking up stairs	0	1	2	3	4	5
6.	While eating	0	1	2	3	4	5
7.	Standing up from a chair	0	1	2	3	4	5
8.	Brushing teeth	0	1	2	3	4	5
9.	Shaving and/or brushing hair	0	1	2	3	4	5
10.	Showering/bathing	0	1	2	3	4	5
11.	Dressing	0	1	2	3	4	5
12.	Picking up and straightening	0	1	2	3	4	5

When I do, or if I were to do, the following tasks, I would rate my breathlessness as:

UCSD Shortness of Breath Questionnaire

0	None at all
1	
2	
3	
4	Severe
5	Maximal or unable to do because of breathlessness

13.	Doing dishes	0	1	2	3	4	5
14.	Sweeping /vacuuming	0	1	2	3	4	5
15.	Making bed	0	1	2	3	4	5
16.	Shopping	0	1	2	3	4	5
17.	Doing laundry	0	1	2	3	4	5
18.	Washing car	0	1	2	3	4	5
19.	Mowing lawn	0	1	2	3	4	5
20.	Watering lawn	0	1	2	3	4	5
21.	Sexual activities	0	1	2	3	4	5

How much do these limit you in your daily life?

22.	Shortness of breath	0	1	2	3	4	5
23.	Fear of "hurting myself" by overexerting	0	1	2	3	4	5
24.	Fear of shortness of breath	0	1	2	3	4	5

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 ever had a seizure, convulsion, or fit?	☐	☐
Have you had recently muscle weakness or pain?	☐	☐
Have you had recently frequent fevers?	☐	☐
Have you lost weight recently?	☐	☐

Appendix 5

Chest CT protocol for ILD patients

New Patients: Standard requisition specifying "New Patient" will be used.

1. Helical CT Inspiration Normal Dose Radiation.

   Reconstruct a. 1 mm HRCT edge enhancing algorithm, plus

   b. 2.5 mm Lung algorithm and

   c. 2.5 mm Soft tissue algorithm

2. Helical Expiration Low Dose Radiation

   Reconstruct a. 2.5 mm Lung algorithm

Routine Expiratory phase for new patients only.

Prone views only if required, as per the chest radiologist assessment. the excess radiation on all patients does not justify the small proportion of patients who may have dependent GG that could be atelectasis vs mild ILD. Request prone chest CT only in those patients.

Follow-up Patients: Standard requisition specifying "Repeat Patient" used.

Helical CT Inspiration Low Dose Radiation.

Reconstruct a. 1mm HRCT edge enhancing algorithm

b. 2.5 mm Lung algorithm

c. 2.5 mm Soft tissue algorithm

Interpretation of chest CTs will be done as per the new Fleischner's Society Criteria as of February, 2018.

Multidisciplinary case discussions

Patients with Surgical lung biopsy +/- bronchoscopy

Pathologist, Chest Radiologists, Respirologist.

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

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 **prefers 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

	DATE d/m/y
Dyspnea Onset (months/years)	
Dyspnea (MRC)	
UCSD dyspnea	
Orthopnea/**P**ND/**l**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	
PFT results	
FVC (N/%)	/
TLC (N/%)	/
Dlco (N/%)	/
6MWT (m/%)	/
6MWT (SaO_2 start/end)	/

Echo date_____ LVEF_____ PAP_____

other: _____

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: _____

CHEST CT Date_____ Report: UIP/possible UIP/Inconsistent/Other_____

LAB Date_____ RF_____ CCP ab____ ANA_____ DsDNA____ ENA_____ ANCA_____

Bronch Date_____ Path_____ BAL Results: N____ L____ E____ M____

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)						
UCSD dyspnea						
Orthopnea/**P**ND/**l**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	**Esbriet_____ Ofev_____** **PPI_____ Other_____**	**Esbriet_____ Ofev_____** **PPI_____ Other_____**	**Esbriet_____ Ofev_____** **PPI_____ Other_____**	**Esbriet_____ Ofev_____** **PPI_____ Other_____**	**Esbriet_____ Ofev_____** **PPI_____ Other_____**	**Esbriet_____ 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_____*** ***Referrals _____***	***GERD measures___*** ***O2 l/min_____*** ***Referrals _____***	***GERD measures___*** ***O2 l/min_____*** ***Referrals _____***

Appendix 9

ILD CLINIC PATIENT RECOMMENDATIONS

1) If you have feathers, down or foam in your pillows or mattress change them to anti-allergic types (Delcron or Polyester filled).

2) Avoid birds, dusts, fumes or chemicals that you might have been exposed to.

3) Remove all visible mold or mildew in walls, ceiling or windows (mix 2 tablespoons of liquid chlorine bleach with 1 quart warm water. Sponge or soak stain for 5 to 15 minutes and then rinse). Keep the indoor humidity between 30% and 50%.

4) If instructed to do so, avoid humidifiers, using bath tubs, hot tubs and Jacuzzis.

5) If the physician 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.

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

7) If you were instructed to purchase a pulse oximeter to monitor your blood oxygen levels (oxygen saturation) and you are using oxygen, titrate it to maintain your oxygen saturation at or above 92% at rest and 88% during exertion.

8) 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.

9) Ask your family doctor if your flu and pneumonia vaccines are up to date. We recommend that you receive your flu vaccine annually and a pneumonia vaccine twice in your life time, five years apart.

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

11) 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.

12) 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 next appointment.

13) If you feel that your condition is worsening (worsening shortness of breath or oxygen levels), please call Dr. Moran's office (phone: 613-548-1380) or go to an emergency department -particularly if your oxygen saturation is less than 90% at rest.

Appendix 10

ILD PROGRAM EDUCATIONAL OBJECTIVES

This form is for recognized specialists whose postgraduate medical training program is designed to give them additional expertise but does not lead to additional credentials for practice. The College of Physicians and Surgeons of Ontario (CPSO) requires the submission of a statement of objectives before issuing a postgraduate education certificate of registration for a clinical fellowship appointment.

Trainee Information

Name of Clinical Fellow: _____ _____
First Name *Last Name*

Specialty Certification:

Title of Certification: _____

Country Issuing Certification: _____

General Information

Department Name: Medicine

Division Name (if applicable): Respirology Division

Name of Fellowship: Interstitial Lung Disease (ILD)

Fellowship Site: Kingston Health Sciences Center

Fellowship Start Date: _____ End Date: _____

If re-appointment:
Reappointment Start Date: _____ End Date: _____

Name of Supervisor: Dr. Onofre Moran-Mendoza

Telephone: 613-548-1380 Email: morano@queensu.ca

Fellowship Overview

Please provide a brief statement of the clinical focus and educational purpose of the fellowship:

The answer space below will expand to accept point form or paragraph entries. If this fellowship is a re-appointment, please describe the clinical focus and educational of the re-appointment only.

DEFINITION

The Interstitial Lung Diseases (ILD) Fellowship is a dedicated training within the Adult Respirology medical subspecialty, concerned with the study, diagnosis and management of Interstitial Lung Diseases.

GOALS

Upon completion of the ILD Fellowship training, a resident is expected to be competent in the assessment and management of Interstitial Lung Diseases.

The resident will develop an in-depth understanding of the most common Interstitial Lung Diseases affecting our population. The resident will acquire the clinical skills to treat and counsel patients with the most common Interstitial Lung Diseases; this will include primary and secondary prevention, treatment and pulmonary rehabilitation. Among others, the professional attributes to be demonstrated and developed will include responsibility, intellectual curiosity, self-appraisal, compassion, and a commitment to continuing medical education. The physician specializing in adult Respirology will also learn to appraise the medical literature critically and to design and carry out research projects.

Interstitial Lung Diseases Fellowship Objectives: CanMEDS Roles

Where applicable, please provide objective (s) for each of the following:

The answer space below will expand to accept point form or paragraph entries; enter "N/A" if individual CanMEDS role is not applicable.

At the completion of ILD Fellowship training, the resident will have acquired the following competencies and will function effectively as a:

1. **Medical Expert**

   As Medical Experts, physicians integrate all of the CanMEDS Roles, applying medical knowledge, clinical skills, and professional attitudes in their provision of patient-centered care. Medical Expert is the central physician Role in the CanMEDS framework.

   A Fellow in Interstitial Lung Diseases (ILD) will be able to function effectively as consultant, integrating all of the CanMEDS Roles to provide optimal, ethical and patient-centered medical care.

   Specifically a Fellow in ILD should be able to:

   a. Achieve sufficient and updated knowledge and clinical skills to properly approach and manage patients with Interstitial Lung Diseases.
   b. Perform a consultation, including a complete and appropriate assessment of a patient with a suspected ILD; and to provide a presentation of well-documented assessments and recommendations in written and/or verbal form in response to a request from another health care professional.
   c. Know and apply current ILD guidelines.

d. Demonstrate proficient and appropriate use of up-to-date diagnostic and therapeutic options in patients with ILD.
e. Correctly order and interpret the results of diagnostic investigations in patients with ILD, including imaging studies, blood work, bronchoscopy, and lung biopsy.
f. Know and identify the characteristic radiological and pathological features of different ILD.
g. Appropriately order, perform and interpret results from Bronchoscopy; including bronchoalveolar lavage and transbronchial, when performed in patients with ILD.
h. Know when to refer patients with ILD to transplant, and what outcomes to expect.
i. Seek and obtain appropriate consultation from other health care professionals for patients with ILD.

2. Communicator

As Communicators, physicians effectively facilitate the doctor-patient relationship and the dynamic exchanges that occur before, during, and after the medical encounter.

A Fellow in Interstitial Lung Diseases will be able to develop rapport, trust, and ethical therapeutic relationships with patients and families.

Specifically a Fellow in ILD should be able to:

a) Be an effective communicator and convey accurately diagnostic and management plans to patients with ILD and to their relatives.
b) Engage patients with ILD, their families and relevant health care professionals in shared decision making to develop a plan of care.
c) Encourage questions, discussions and interaction in the clinical encounter to ensure that the medical options offered to patients with ILD –including the goals of care at the end of life-, are in keeping with the patients' and relatives' preferences and values.
d) Describe and apply the bioethical principles in obtaining informed consent for diagnostic or therapeutic procedures in patients with ILD.
e) Respect patient confidentiality, privacy and autonomy.
f) Convey effective oral and written information about the medical encounters.

3. Collaborator

As Collaborators, physicians effectively work within a healthcare team to achieve optimal patient care.

A Fellow in Interstitial Lung Diseases will be able to participate effectively and appropriately in an interprofessional health care team.

Specifically a Fellow in ILD should be able to:

a) Work with others to assess, plan, provide and integrate care for individual patients (or groups of patients) with ILD.
b) Develop a care plan for the patient with ILD in collaboration with members of the interprofessional team.
c) Coordinate and present cases at the monthly ILD rounds.

d) Work with others to assess, plan, provide and review other tasks, such as research problems, educational work, program review or administrative responsibilities.
e) Participate in interprofessional team meetings, demonstrating the ability to accept, consider and respect the opinion of other team members while contributing specific expertise.
f) Enter into interdependent relationships with other professions for the provision of quality care to patients with ILD.
g) Demonstrate leadership in a health care team, as appropriate.

4. Manager

As Managers, physicians are integral participants in healthcare organizations, organizing sustainable practices, making decisions about allocating resources, and contributing to the effectiveness of the healthcare system.

A Fellow in Interstitial Lung Diseases will be able to participate in activities that contribute to the effectiveness of their health care organizations and systems, as well as to serve in administration and leadership roles as appropriate.

Specifically a Fellow in ILD should be able to:

a) Work collaboratively with others in their organizations.
b) Implement processes to ensure practice improvement.
c) Employ information technology appropriately for patient care.
d) Lead or participate effectively in clinical rounds and meetings.

5. Health Advocate

As Health Advocates, physicians responsibly use their expertise and influence to advance the health and well-being of individual patients, communities, and populations.

A Fellow in Interstitial Lung Diseases will be able to respond to individual patient health needs and issues as part of patient care and identify the determinants of health of patients with ILD.

Specifically a Fellow in ILD should be able to:

a) Identify the health needs of individual patients with ILD.
b) Identify the determinants of health of patients with ILD, including barriers to access to care and resources.
c) Advocate for ILD patients with obstacles to access diagnostic and therapeutic resources.

6. Scholar

As Scholars, physicians demonstrate a lifelong commitment to reflective learning, as well as the creation, dissemination, application and translation of medical knowledge.

A Fellow in Interstitial Lung Diseases will be able to maintain and enhance professional activities through ongoing learning, as well as to critically evaluate medical information and to be able to generate new knowledge.

Specifically a Fellow in ILD should be able to:

a) Create and maintain an active and ongoing learning process.
b) Integrate new learning into practice.
c) Critically appraise obtained evidence regarding ILD.
d) Integrate critical appraisal conclusions into the clinical care of patients with ILD.
e) Contribute to the development and dissemination of new knowledge and practices in ILD.

7. Professional

As Professionals, physicians are committed to the health and well-being of individuals and society through ethical practice, profession-led regulation, and high personal standards of behaviour.

A Fellow in Interstitial Lung Diseases will be able to demonstrate a commitment to their patients, profession, and society through ethical practice

Specifically a Fellow in ILD should be able to:

a) Exhibit appropriate professional behaviors in practice, including honesty, integrity, commitment, compassion, respect and altruism.
b) Demonstrate a commitment to delivering the highest quality care and maintenance of competence.
c) Maintain appropriate professional relations with patients, colleagues and other health care team members.
d) Recognize the principles and limits of patient confidentiality as defined by professional practice standards and the law.
e) Fulfill the regulatory and legal obligations required of current practice.

EVALUATION:

Evaluations will be done quarterly, addressing the CANMED roles and using the In-Training Respirology Evaluation Form (attached). In addition, the specific evaluation form for Saudi Trainees will be used, as requested by the Saudi Bureau. There will be formative evaluations and feedback provided to the ILD Fellow on a regular bases during clinics, after reviewing every patient, as well as after the monthly ILD round presentation. These encounters will be the sources of information for the evaluations.

Additional specific objectives: (Optional)

Onofre Moran, M.D. MSc, PhD, FRCPC
Director, Interstitial Lung Disease Program,
Queen's University.