Interstitial Lung Disease: Not all SOB is COPD!!

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Relationships with Commercial Interests:

- **Grants/Research Support:** none
- **Speakers bureau/Honoraria:** Astra Zeneca, Boehringer-Ingelheim, ICEBM, MD Briefcase
- **Consulting Fees:** none
- **Other**: Director Clinical teaching rounds, Mount Sinai Hospital
  CTS cannabis guidelines

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Disclosure of Commercial Support

- This program has received no financial or in kind support
- Potential for conflict of Interest
  - I have received consultancy fees and speaker fees from a number of respiratory organizations
  - Many respiratory products will be mentionned in this talk (generic names only)

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CFPC CoI Templates: Slide 3

Mitigating Potential Bias

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I have independently reviewed all products to be presented And have attempted to mention all products currently Available in Canada. I will discuss all diagnostic and treatment options for respiratory care

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Our Case: Fred W.

- 65 year old male
- 35 pack year smoker; quit 15 years ago
- One treatment with prednisone last year when he couldn't breathe
- Meds; tiotropium and salbutamol
- Ran out of puffers 2 weeks ago
- Dry cough almost all the time

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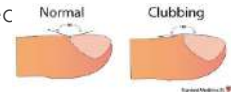
The exam

- Short of breath sitting in waiting room and walks slowly to your office, stopping twice
- Lips and fingertips a little blue by the time he sits on exam table
- Bp 110/75 p 105 r 32
- O2 sat 88%
- Chest- decreased a/e ?crack
- Fingers and toes clubbed



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More history

- many jobs, including in a chemical processing plant and as a mechanic for locomotives
- Last 10 years worked as office manager at locomotive plant
- Retired age 62 because "it was time"
- Golfed until last year; always used a cart.
- Drives a car; goes out only to help wife with shopping

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What do you think?

- Asthma?
- COPD?
- ACOS?
- Lung Cancer?
- Interstitial lung disease?
- CHF?

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?COPD

- Sob, cough, smoking history;
  - First thought; COPD!
  - Treatment; add LABA to current treatment and follow up in 6 months…

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Six months later…..

- Two exacerbations treated with prednisone and antibiotics
- SOB with all activity
- Barely leaves house
- Wife has to help him bathe and dress

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Exam

- Holds on to his wife when walking into office
- BP 110/78, P 115, R 36
- Wt loss 10 pounds since last visit
- O2 sat 88%
- Chest- ↓air entry, bibasilar crackles
- Clubbing as previous, fingertips blue

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Workup

- Spirometry
- Mrc dyspnea scale / CAT
- CXR
- CT scan

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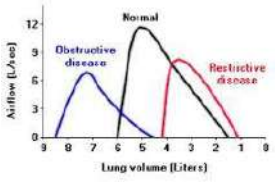
Results for Fred

- Spirometry;
  - FEV1= 1.65 (70%)
  - FVC= 2.15 (60%)
  - FEV1/FVC= 0.77
- MRC = 4/5



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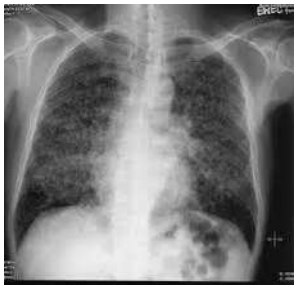
Results;

- CXR



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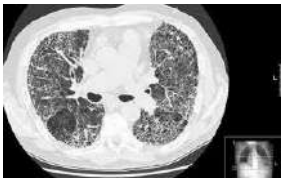
Further workup

- HRCT
  - UIP pattern on HRCT is indicative of a definite diagnosis of IPF



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Interstitial Lung disease

Characterized by

-**progressive** scarring of the lung tissue (**fibrosis)**

- dyspnea and hypoxia.

- categorized into known and unknown causes
- scarring is generally irreversible

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Interstitial lung disease; causes

- **autoimmune or rheumatologic diseases**
  - Lupus, rheumatoid arthritis, sarcoidosis, scleroderma
- **occupational and organic exposures**;
  - coal dust, farmers lung, asbestos, iron dust (siderosis), silica
  - Molds, gases, fumes
- **medications**
  - Nitrofurantoin, sulfonamides, bleomycin, amiodarone, methotrexate, gold, infliximab, and etaercept
- **Radiation**
- **Unknown (idiopathic)**

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Idiopathic Pulmonary Fibrosis

- Chronic, progressive, fibrosing, interstitial pneumonia of unknown cause
- Occurs in adults, usually > 60 yo
- Limited to the lungs
- Disease likely more fibrotic than inflammatory
- Prognosis poor (20-30% 5 yr survival)

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Idiopathic Pulmonary Fibrosis

- Disease progression usually insidious until late stages
- Rate of decline in FVC =150-200ml/yr
- Course of disease often unpredictable
- Symptoms often mistaken for COPD
- Frequent co morbid conditions include COPD and CHF
- Usually classified as mild, moderate, severe
- Oxygen often required in severe disease (more with exercise)

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IPF; Clinical presentation

- Exertional dyspnea (insidious onset)
- Cough (usually dry); Intractable
- Bibasilar inspiratory crackles"velcro-like"
- Finger clubbing ((25-50%)
- Acute exacerbations uncommon
  - Hypoxia with new infiltrates of unclear etiology
  - May be initial presentation

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Epidemiology

- Rare disease; difficult to study
- Prevalence 0.7-63.0/100,000[1]
- Canada : No accurate prevalence data[2]
  - 3,000-4,000 new cases/year
  - Estimated 5,000-10,000 patients

1. Ley B, Collard HR. *Clin Epidemiol* 2013;5:483-92. 2. Kingsberg S et al. *Int J Womens Health* 2010;1:105-11.

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Epidemiology

- 97% of Canadian patients with IPF initially present to their family doctor, only **15%** of FP's suggested IPF as possible diagnosis
- Ave time to diagnosis; 20 months
- 32% of patients received another diagnosis (15% received > 3!)
  - www.canadianpulmonaryfibrosis.ca

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Risk factors

- Cigarette smoking[1,2]
  - particularly if smoking history of >20 pack-years
  - Note: IPF should be considered even if smoking history is absent
- Environmental exposures[3]
  - Farming, livestock, raising birds, vegetable dust/animal dust, metal dust, stone cutting/polishing, hairdressing

1. Raghu G et al. *Am J Respir Crit Care Med* 2011;183(6):788-824. 2 Baumgartner KB et al. *Am J Respir Crit Care Med* 1997;155(1):242-248. 3. Baumgartner KB et al. *Am J Epidemiol* 2000;152(4):307-315.

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Risk factors (contd.)

- Microbial agents[1]
  - Viruses may play a role in IPF initiation, progression, and exacerbations
  - Role of bacteria is less clear
- Gastroesophageal reflux[2,3]
  - Micro-aspiration of gastric contents causes repetitive lung injury and pulmonary fibrosis in susceptible individuals
- Genetic mutations
  - Associated with 15% of familial IPF

1. Molyneaux PL, Maher TM. *Eur Respir Rev* 2013;22(129):376-381. 2. Ley B, Collard HR. *Clin Epidemiol* 2013;5:483-92. 3. Raghu G et al. *Euro Respir J* 2006;27(1):136-142.

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Disease Severity

- Mild:
  - asymptomatic or mild cough
  - dyspnea with substantial exertion
- Moderate:
  - dyspnea on moderate exertion
  - non-productive cough,
  - mild to moderate PFT abnormalities (↓FVC 50-70%pred,↓DLCO 45-60% pred)
  - desaturation with effort
- Severe:
  - dyspnea on mild exertion
  - Require O2 at rest and/or exertion
  - PFT changes (FVC<50%pred,DLCO<50%pred)

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Natural history

- Progressive decline in pulmonary function
- Course: Mostly slow and gradual progression over many years
  - Some patients:
    - Rapid progression
    - Periods of relative stability and acute decline
- Most frequent cause of death: Respiratory failure

Ley B et al. *Am J Respir Crit Care Med* 2011;183(4):431-440.

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Natural history (contd.)

- Median survival: 2-3 years
- Factors for shorter survival
  - Older age
  - Smoking history
  - Lower BMI
  - More severe physiologic impairment
  - More radiologic disease
  - Other complications/conditions: PH, emphysema, malignancy

Ley B et al. *Am J Respir Crit Care Med* 2011;183(4):431-440.

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Natural History ;comorbidities

- Hypoxemia
- Pulmonary hypertension
- Thrombo-embolic disease
- COPD
- Heart failure
- OSA
- Depression

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Management

- Nutrition; dietary supplements
- Exercise
- Vaccines; flu vaccine, pneumonia vaccine
- Oxygen?
- Prevention of reflux and recurrent microaspiration may slow disease progression
- Referral to respirology for further investigations, including bronchoscopy and biopsy

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Medications for IPF

- Corticosteroids, Immunosuppressants –
  - Several trials and lots of clinical experience; lack of efficacy and ↑adverse events (including hospitalization and death)
- Colchicine, interferon, endothelin receptor antagonists, N acetyl cysteine
  - no benefit

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Medications for IPF

- IPF is a fibrosing disease (NOT inflammatory)
- No cure, BUT 2 drugs (nintedanib and pirphenidone) may slow disease progression
- Indicated for MILD to MODERATE IPF without underlying liver disease

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Medications for IPF

- Pirfenidone– antifibrotic agent
  - May decrease decline in FVC
  - Indicated in mild to moderate IPF
  - Side effects; rash, photosensitivity, nausea, diarrhea, abd discomfort, dyspepsia, anorexia, fatigue
  - ↑LFT's, interaction CYP1A2 inhibitors
- Nintedanib- receptor blocker for tyrosine kinase
  - May reduce rate of decline in lung function
  - Appears to slow rate of disease progression
  - Side effects; ↑LFT's, ↑bleeding (on AC), CYP3A4 inhibitors
  - Diarrhea, nausea, vomiting

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Our Case, six months later

- Resp consult; biopsy compatible with IPF
- Pulm rehab 2 months ago
- Home O2 for past 3 months
- Improved exercise tolerance
- more autonomy
- Trial Pirfenidone; severe rash; discontinued
- Started on nintedanib 2 months ago
  - Initial diarrhea; improved with dose reduction
  - Monthly LFT's (ALT,AST,Bilirubin) ok

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Questions?

- http://www.canadianpulmonaryfibrosis.ca
- Lung.ca
- Cfpc.ca
- Slevitz.sinai@ssss.gouv.qc.ca

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