Lung Health Community of Practice Series 1

Pulmonary Fibrosis

Project ECHO
Palliative Care - Canada
BY Pallium Canada

Facilitator: Diana Vincze, Pallium Canada
Presenters: Dr. Alan Kaplan and Geneviève Lalumière
Date: 28 June 2024

Territorial Honouring

The Palliative Care ECHO Project

The Palliative Care ECHO Project is a 5-year national initiative to cultivate communities of practice and establish continuous professional development among health care providers across Canada who care for patients with life-limiting illness.

Stay connected: www.echopalliative.com

The Palliative Care ECHO Project is supported by a financial contribution from Health Canada. The views expressed herein do not necessarily represent the views of Health Canada.

Health Canada Santé Canada

Project ECHO Palliative Care - Canada BY Pallium Canada

LEAP Lung

- Learn the essentials for providing a palliative care approach.
- Ideal for any health care professional (e.g. physician, nurse, pharmacist, social worker, etc.) who provide care to patients with advanced lung diseases.

- **Key features:**
  - Created and reviewed by Canadian experts
  - Evidence-based
  - Regularly updated and approved
  - Practical, case-based
  - Accredited

Pallium Canada

LEAP LUNG

Learn more about the course and topics covered by visiting

https://www.pallium.ca/course/leap-lung/

Introductions

Facilitator
Diana Vincze
Palliative Care ECHO Project Manager, Pallium Canada

Panelists
Danielle Hill RRT, CRE, CSFI
Respiratory Therapist, Arnprior And District Family Health Team

Jody Hamilton, BSW, MSW
Director Community Programs & Partnerships, Lung Health Foundation

Dr. Joshua Wald, MD, FRCPC (respirologist)
Associate Professor

Presenters
Dr. Alan Kaplan, MD CCFP(EM) FCFP CPC(HC)
Chairperson, Family Physician Airways Group of Canada
Clinical Lecturer, Dept of Family and Community Medicine, University of Toronto

Geneviève Lalumière, BScN, RN MN
Clinical Nurse Specialist and Coordinator Regional Palliative Consultation Team, Bruyère Continuing Care

Disclosure

Relationship with Financial Sponsors:

Pallium Canada

- Not-for-profit
- Funded by Health Canada
- Boehringer Ingelheim supports Pallium Canada through an in-kind grant to expand interprofessional education in palliative care.

Disclosure

This program has received financial support from:

- Health Canada in the form of a contribution program
- Pallium Canada generates funds to support operations and R&D from Pallium Pocketbook sales and course registration fees
- An educational grant or in-kind resources from Boehringer Ingelheim.

Facilitator/ Presenter/ Panelists:

- **Diana Vincze**: Palliative Care ECHO Project Manager at Pallium Canada.
- **Geneviève Lalumière:** Nothing to disclose
- **Dr. Alan Kaplan:** Speaking Engagements/Honoraria/Consulting fees: ALK, Astra Zeneca, Boehringer Ingelheim,, Covis, Eisai, GSK, Idorsia, Pfizer, Moderna, NovoNordisk, Sanofi, Teva, Trudell, Valeo. Educational companies: MD Briefcase, PeerView, Respiplus.
- **Jody Hamilton:** Nothing to disclose
- **Danielle Hill:** Speaker/Honoraria fees from GSK and AstraZeneca
- **Dr. Joshua Wald:** Speaking fees and honoraria from GSK, AstraZeneca, Canadian Institute for the transfer of knowledge (CITE) and the lung health foundation.

Disclosure

Mitigating Potential Biases:

- The scientific planning committee had complete independent control over the development of program content

Welcome and Reminders

- Please introduce yourself in the chat!
- Your microphones are muted. There will be time during this session for questions and discussion.
- You are also welcome to use the Q&A function to ask questions, but also feel free to raise your hand!
- This session is being recorded and will be emailed to registrants within the next week.
- Remember not to disclose any Personal Health Information (PHI) during the session.
- Each session has been approved for 1.0 CSRT CPD credit by the Canadian Society of Respiratory Therapists (CSRT).
- This event is also an Accredited Group Learning Activity through the Royal College of Physicians and Surgeons of Canada. You may claim a maximum of **5.00 hours.**

Objectives of this Series

After participating in this program, participants will be able to:

- Describe what others have done to integrate palliative care services into their practice.
- Share knowledge and experience with their peers.
- Increase their knowledge and comfort around integrating a palliative care approach for their patients with advanced lung disease.

Overview of Topics

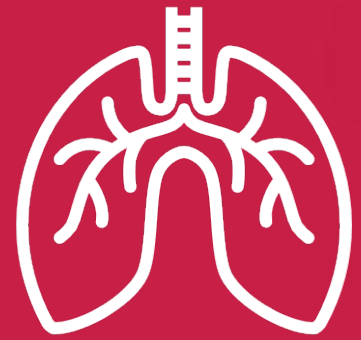
Session #	Session title	Date/ Time
Session 1	Palliative care in advanced respiratory illnesses	February 28, 2024 from 12-1pm ET
Session 2	COPD Management	May 1, 2024 from 12-1pm ET
Session 3	Pulmonary Fibrosis	June 28, 2024 from 12-1pm ET
Session 4	Symptom management in advanced respiratory illnesses	September 18, 2024 from 12-1pm ET
Session 5	Psychological distress and depression	November 27, 2024 from 12-1pm ET

Objectives of this Session

After participating in this session, participants will be able to:

- Understand the Pathophysiology and Progression of Pulmonary Fibrosis
- Optimize Pharmacological and Non-Pharmacological Interventions
- Integrate Palliative Care Principles into Pulmonary Fibrosis Management
- Promote Interdisciplinary Collaboration for Holistic Patient Care
- Manage Pulmonary Fibrosis at EOL

Idiopathic Pulmonary Fibrosis



Presenter Disclosure

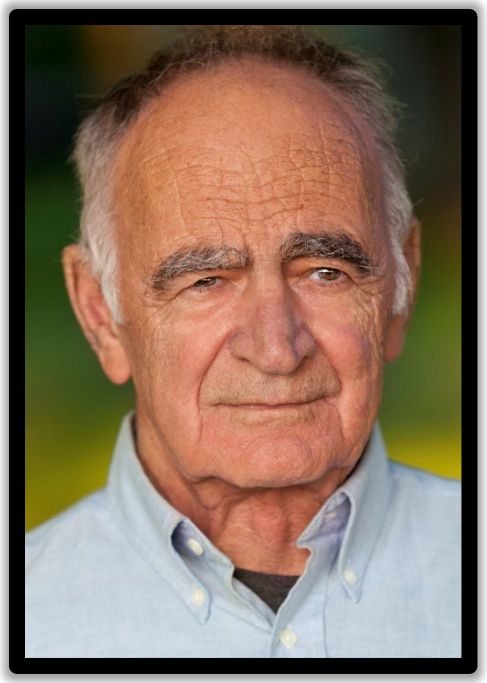
- Faculty: Alan Kaplan MD CCFP(EM) FCFP CPC(HC)
- Chairperson, FPAGC
- Community Chronic Pain consultant
- Community Respiratory consultant

- Relationship With Commercial Interests:
- Multiple companies in Respiratory and Pain worlds including Spectrum re Cannabis

- Potential for Conflict of Interest:
- Past consultant for BI which makes IPF meds, however NONE for >5 years

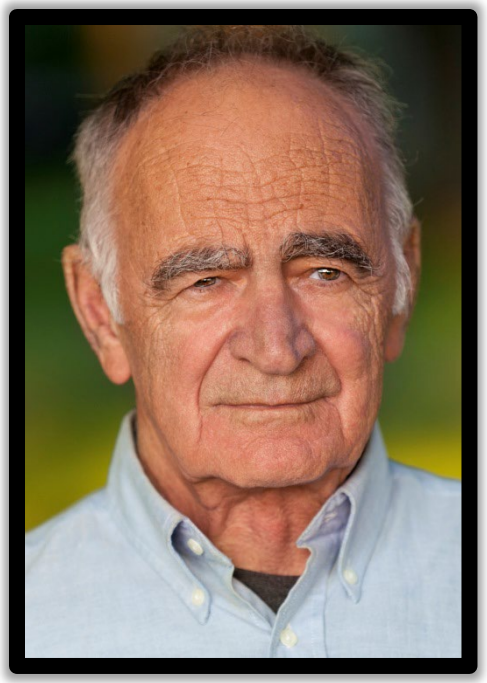
Meet the patient:
Eric: 65-year-old male

- Non-productive cough – 1 year
- Shortness of breath – 1 month
- Former 20 pack-year smoker; quit 2 years ago
- Being treated by PCP for COPD
- Despite inhaled dual long-acting bronchodilator dyspnea becoming worse
- Cough not responding to antitussive meds



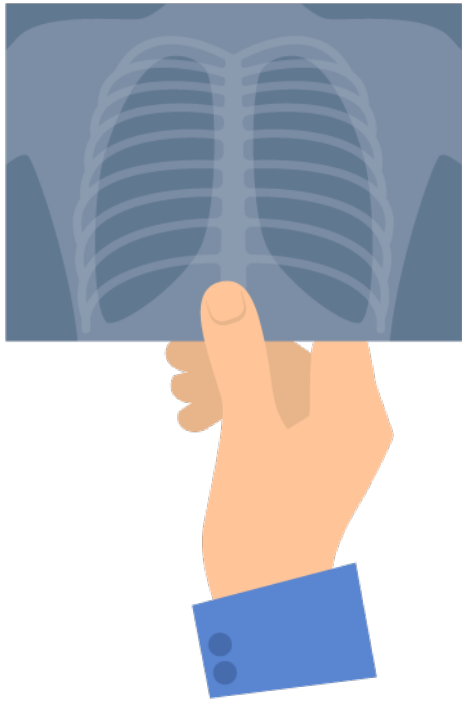
Meet the patient:
Eric: 65-year-old male

- Auscultation: Late inspiratory crackles
- Examination otherwise unremarkable
- Chest X-ray ordered: Bilateral, peripheral interstitial opacities
- Spirometry:
  - FEV_1/FVC ratio: 84% of predicted
  - FEV1 87% predicted
  - FVC 71% predicted



When to consider ILD?

- Patients with presumed COPD/HF, not benefiting from treatment
- Subtle abnormalities on chest X-ray (largely normal in COPD)
- Middle-aged/elderly patients with unexplained chronic exertional dyspnea or chronic cough
- No obstruction on Spirometry!



Adpated from . Zibrak JD et al. *NPJ Prim Care Respir Med*. 2014;24:14054.

Drug-induced ILD

- Common causes:
  - Cancer chemotherapy, including methotrexate
  - Nitrofurantoin
  - Amiodarone

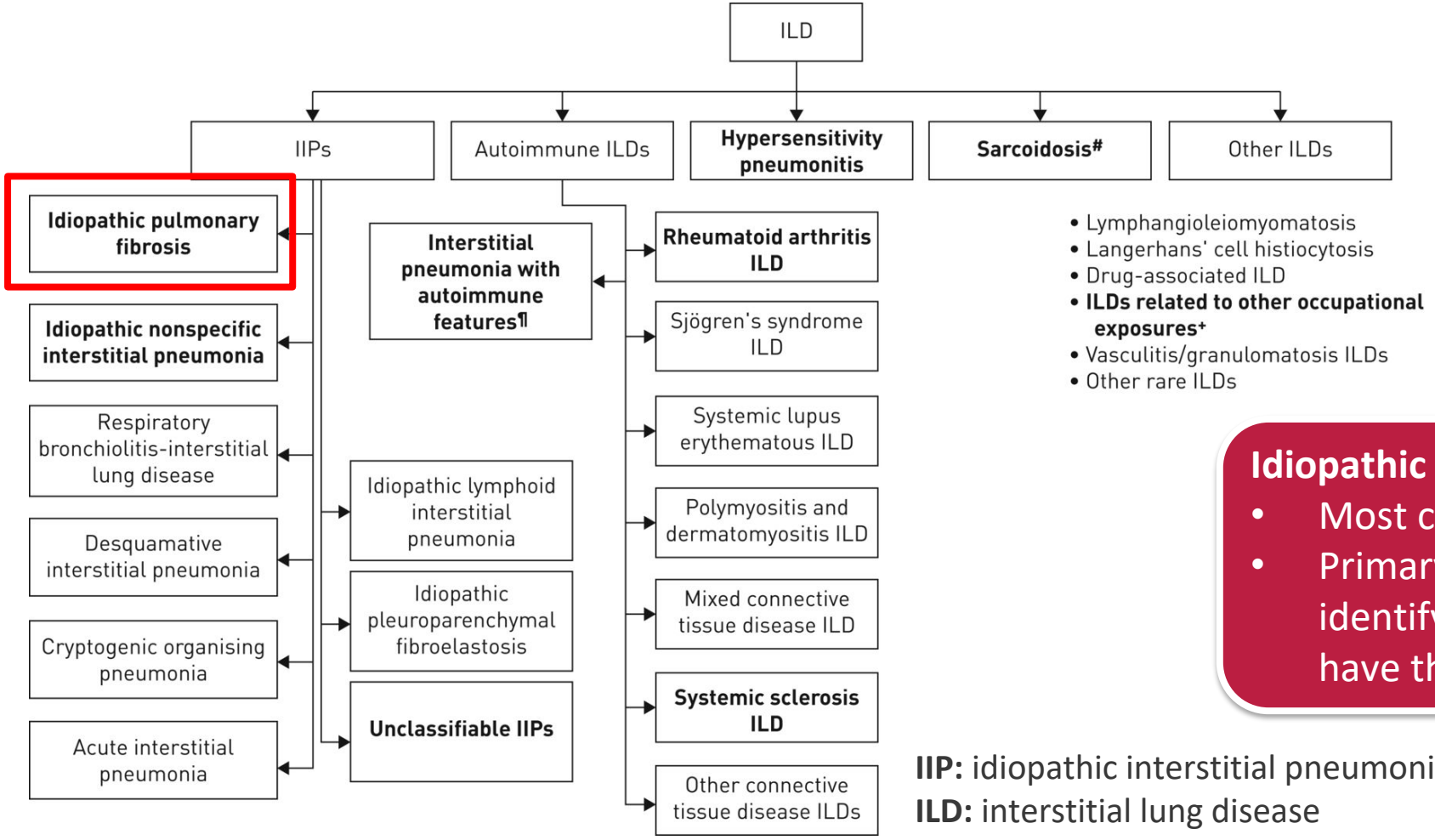
Schwaiblmair M, Behr W, Haeckel T, Märkl B, Foerg W, Berghaus T. Drug Induced Interstitial Lung Disease. Open Respir Med J. 2012;6:63-74. doi:10.2174/1874306401206010063

IPF vs. COPD

	IPF	COPD
Physical examination		
Inspiratory crackles	Late, velcro	Early
Wheezing	✗	✓
Finger clubbing	+/-	✗
Hyperinflation	✗	✓
Spirometry		
Reduction in TLC	✓	✗
Reduced FEV1/FVC ratio	✗	✓
Others		
Response to bronchodilators	✗	✓
Chest X-ray	Specific patterns of abnormality, may be Normal	Largely normal, occ hyperinflation

Zibrak JD. *NPJ Prim Care Respir Med*. 2014;24:14054.

Categories of Interstitial Lung Disease (ILD)



Idiopathic pulmonary fibrosis (IPF)

- Most common ILD
- Primary care is crucial for identifying patients who may have this condition

IIP: idiopathic interstitial pneumonias
ILD: interstitial lung disease

Bolded ILDs are most likely to have a progressive-fibrosing phenotype

Cottin V, Hirani NA, Hotchkin DL, et al. Presentation, diagnosis and clinical course of the spectrum of progressive-fibrosing interstitial lung diseases. *European Respiratory Review*. 2018;27(150). doi:10.1183/16000617.0076-2018

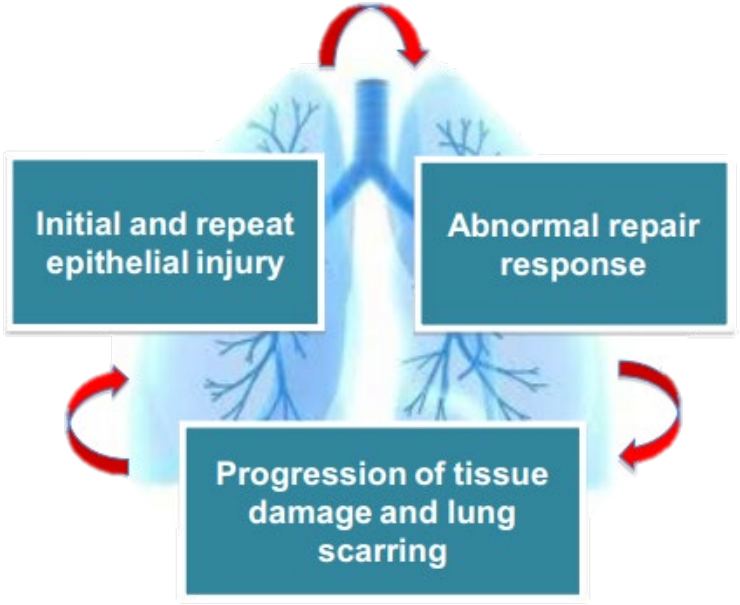
Idiopathic pulmonary fibrosis (IPF)

- 20% of all ILDs; most frequent and severe form of idiopathic interstitial pneumonia (IIP)
- Unknown cause
- Primarily occurs in older adults
- Disease course varies, average survival little better than inoperable lung cancer
- Delayed diagnosis may negatively impact patients

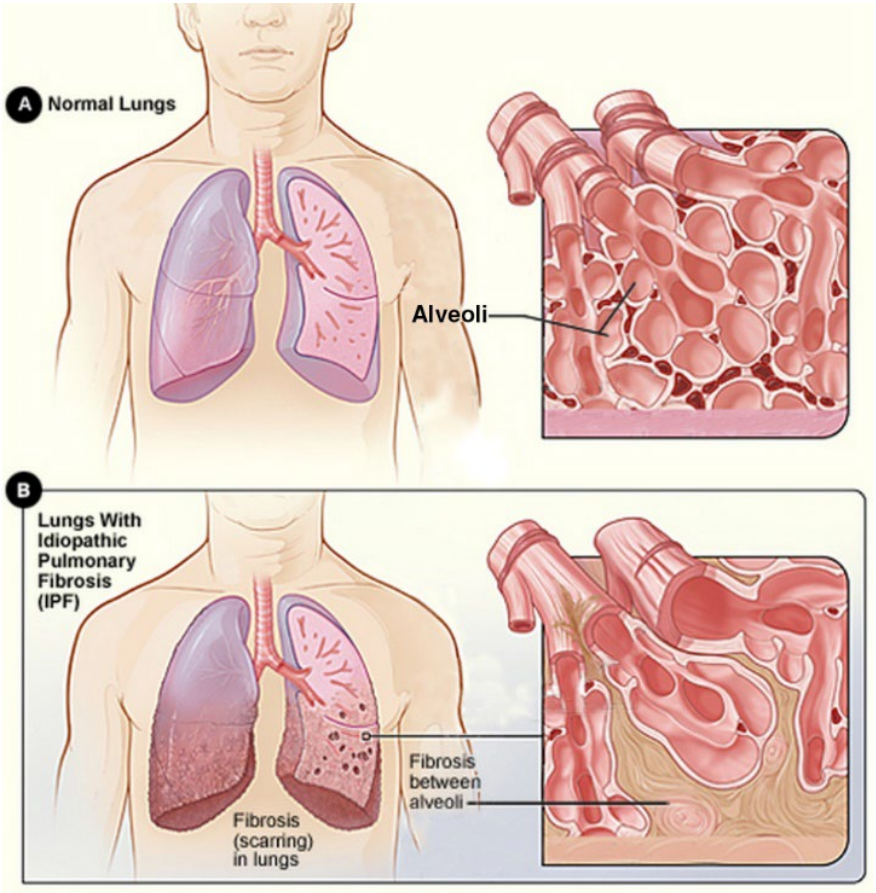
1. Raghu G et al. *Am J Respir Crit Care Med.* 2015;192(2):e3-19. 2 Sgalla G et al. *Respirology.* 2016;21(3):427-37.
3. Antoniou KM et al. *Lancet Respir Med.* 2014;2(1):e1. 4. http://formularyjournal.modernmedicine.com.

IPF: abnormal wound healing leads to irreversible fibrosis

- Three stages of pathogenesis[1]:



- What initiates this process is unknown

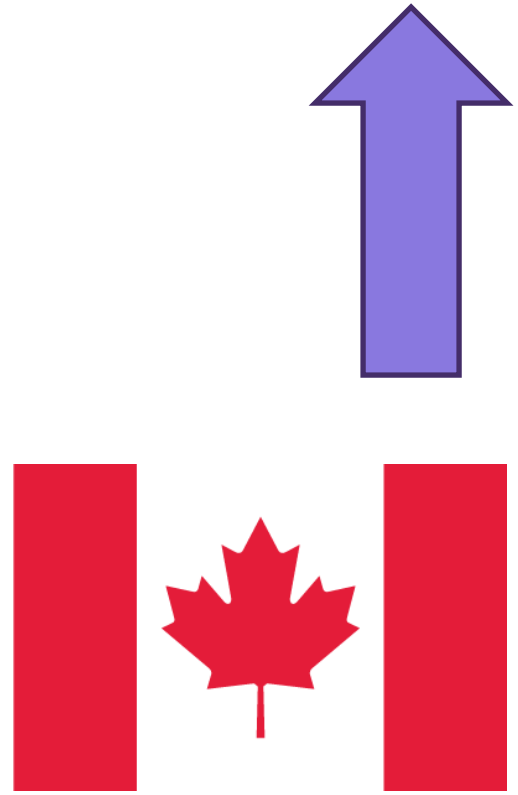


- Fibrosis distinguishes IPF from other ILDs, which are mainly inflammatory[2]
- Distorted alveolar−capillary barrier architecture leads to impaired gas exchange, which limits routine physical activity[3]

1. Travis WD et al. *Am J Respir Crit Care Med*. 20
188(6):733–748.
2. Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824.
3. Meltzer EB, Noble PW. *Orphanet J Rare Dis*. 2008;3:8−22.

High rates of IPF in Canada

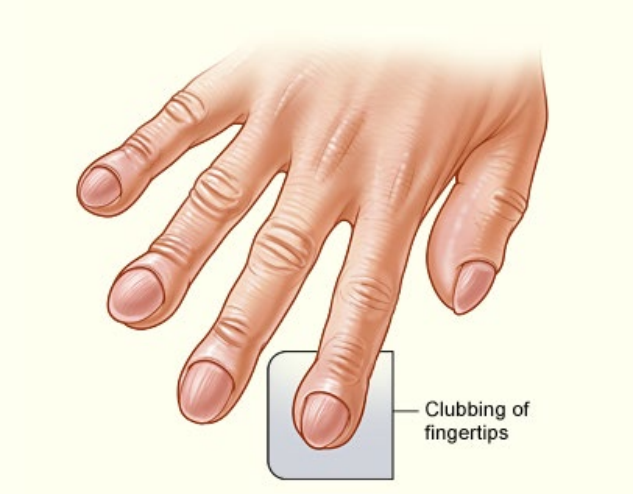
- Published studies:
  - Prevalence 0.7-63.0/100,000 person-years
  - Incidence 0.6-17.4/100,000 person-years
- Higher prevalence/incidence in Canada than in other jurisdictions:
  - Prevalence: 54-115/100,000
  - Incidence: 24-51/100,000



5. Ley B, Collard HR. *Clin Epidemiol.* 2013;5:483-492. 6. Charlene DF, et al. *Am J Respir Crit Care Med.* 2015;191:A2522-A2522.

Common clinical presentation of IPF

- Older age (>60 years)
- Male gender
- Smoking common, in 60−70%
- Symptoms:
  - Progressive exertional dyspnea
  - Non-productive cough
- Signs:
  - Bilateral inspiratory Velcro-like crackles
  - Clubbing of fingers
- Tests:
  - Abnormal pulmonary function test indicating restriction and impaired gas exchange



Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824.

IPF risk factors

- **Smoking**: Strong association, particularly if >20 pack-years
- **GERD**: Microaspiration of gastric contents → repetitive lung injury
- **Genes**: *TERT, TERC*, *SPC, SPA2*, *ELMOD2*, *MUC5B*
- **Viruses**: Role in initiation, progression, exacerbations

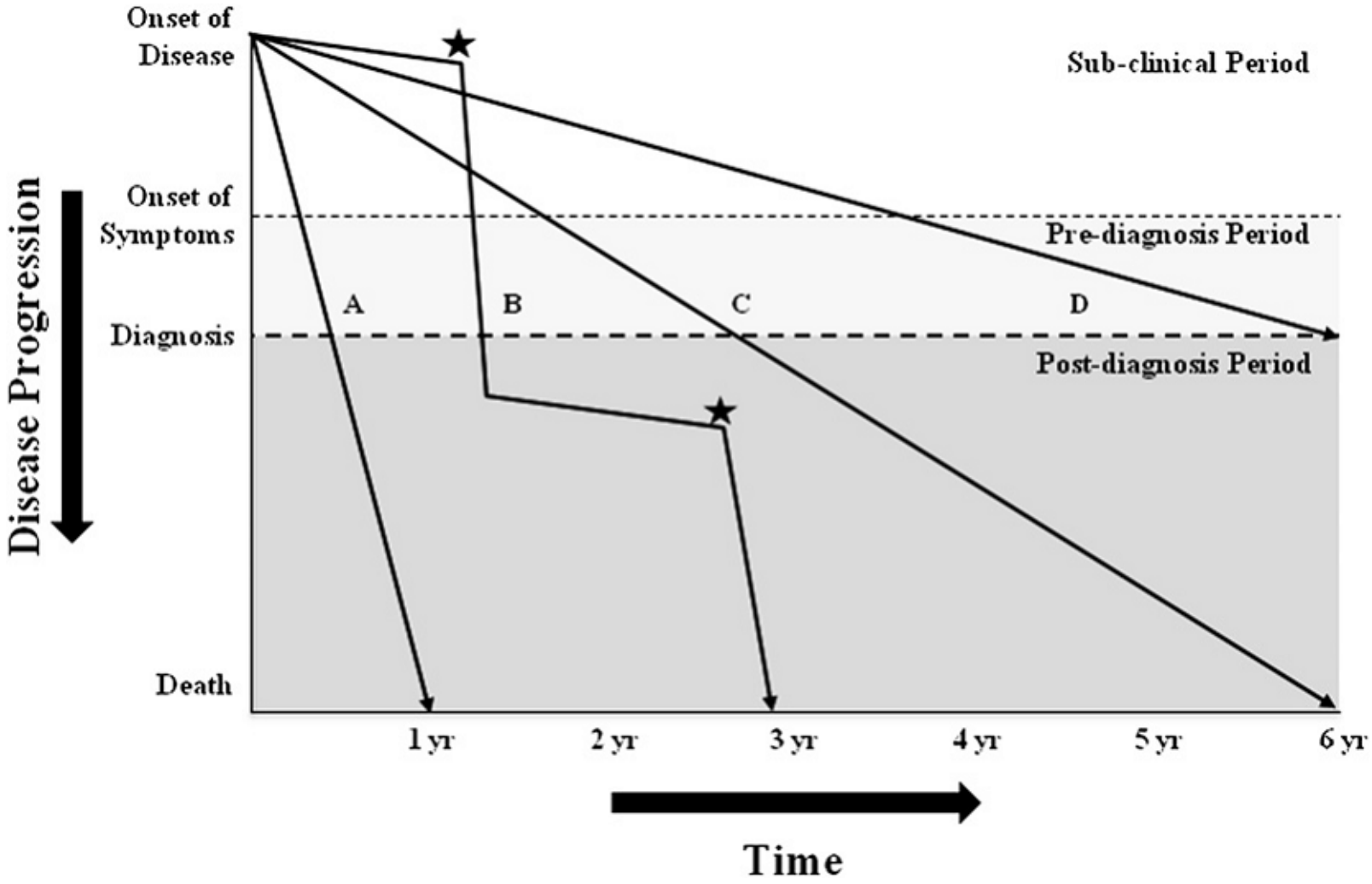
5. Ley B et al. *Clin Epidemiol*. 2013;5:483-492. 7. Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788-824.
8. Baumgartner KB et al. *Am J Respir Crit Care Med*. 1997;155(1):242-248. 9. Molyneaux PL et al. *Eur Respir Rev*. 2013;22(129):376-381. 10. Raghu G et al. *Euro Respir J*. 2006;27(1):136-142.

Natural history of IPF

- Progressive disease - at variable rates
- Median survival from diagnosis: 2-3 years
- Death most frequently from respiratory failure
- **Factors associated with shorter survival**:
  - Older age, smoking history, lower BMI
  - More severe abnormalities on lung function tests and exercise tests,
  - Greater radiologic extent of disease
  - Complications (pulmonary arterial hypertension)

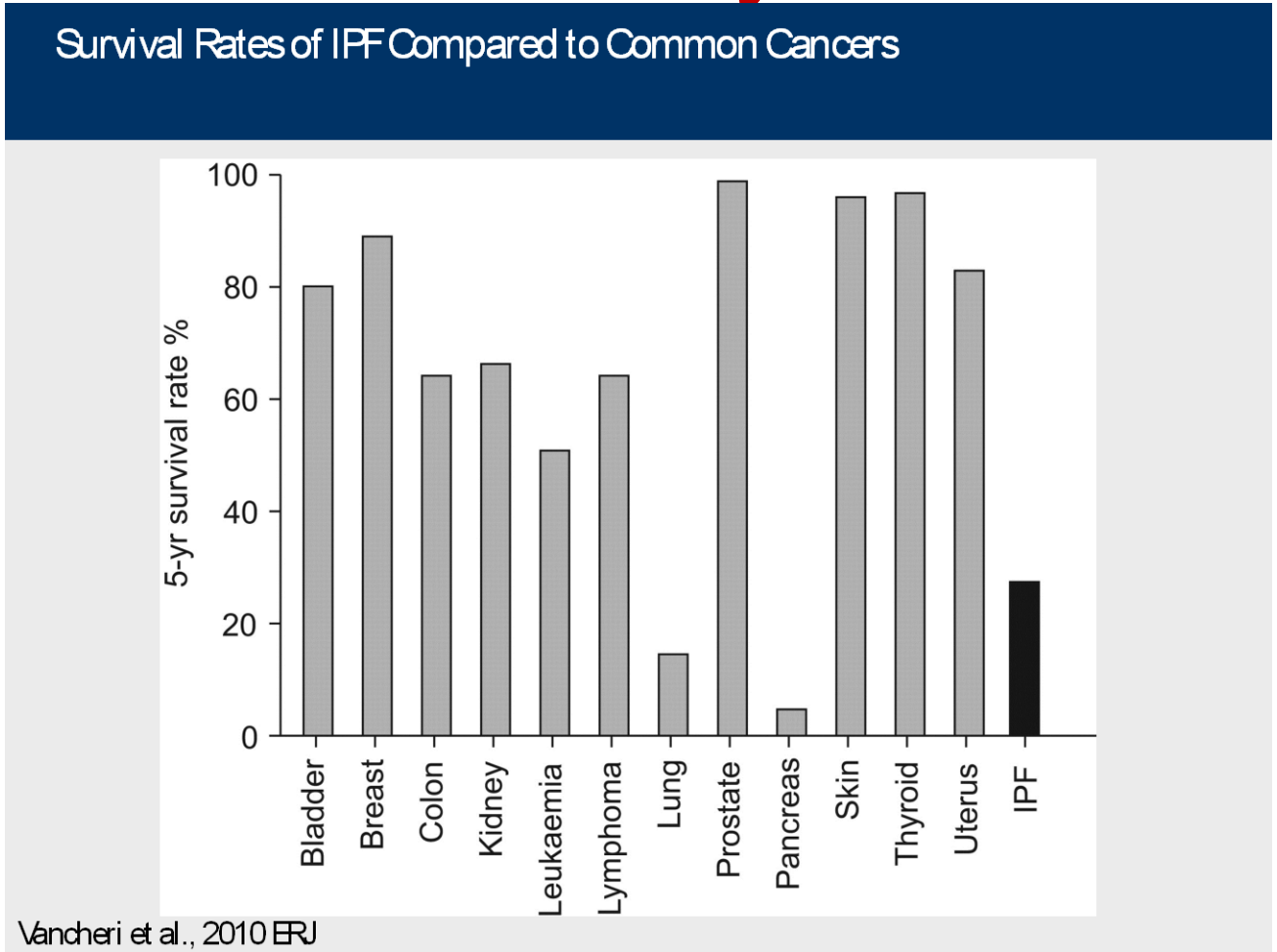
2. Sgalla G et al. *Respirology*. 2016;21(3):427-37. 17. Ley B et al. *Am J Respir Crit Care Med.* 2011;183(4):431-440.

Natural history of IPF



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

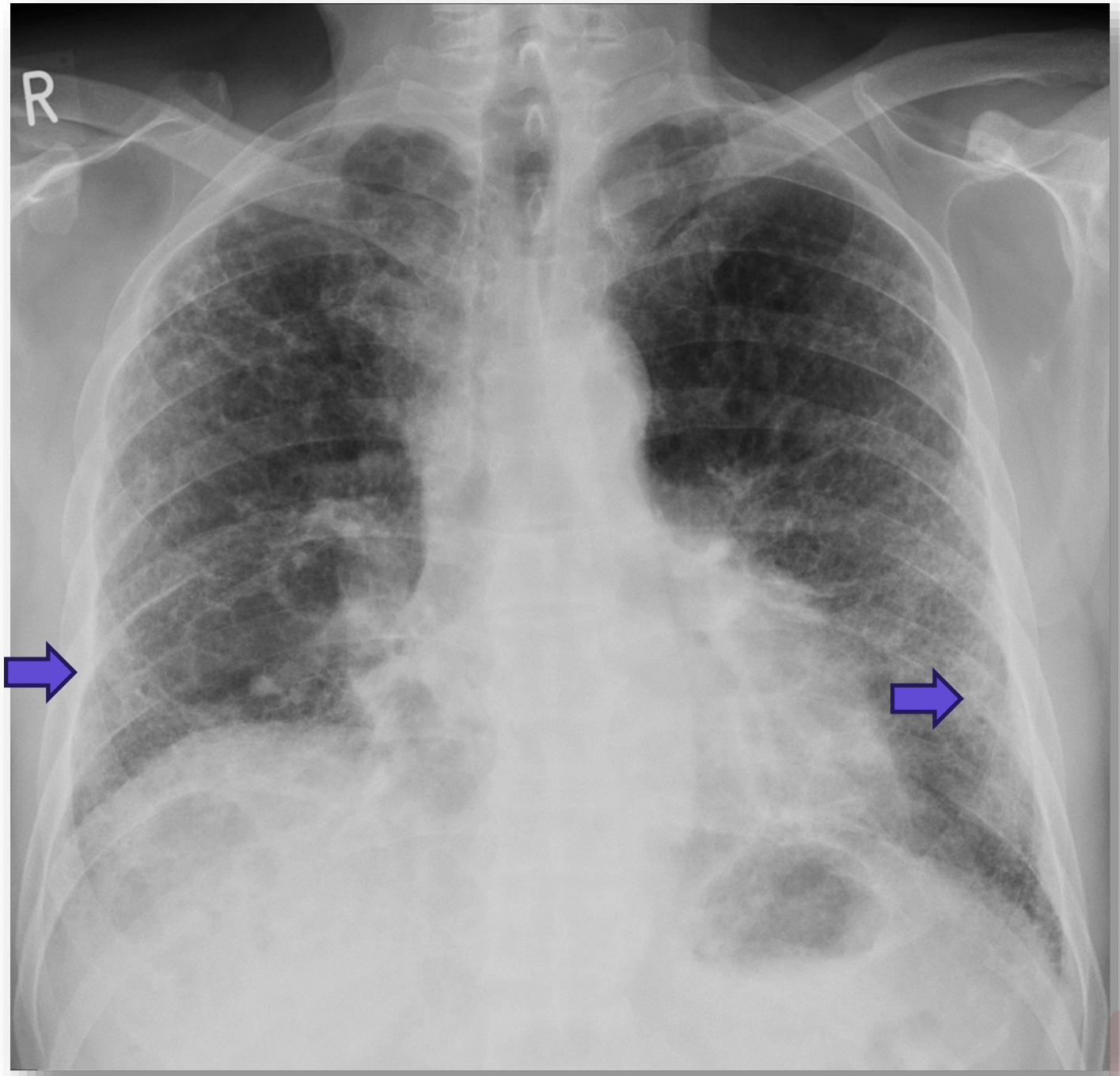
IPF – As lethal as many common cancers



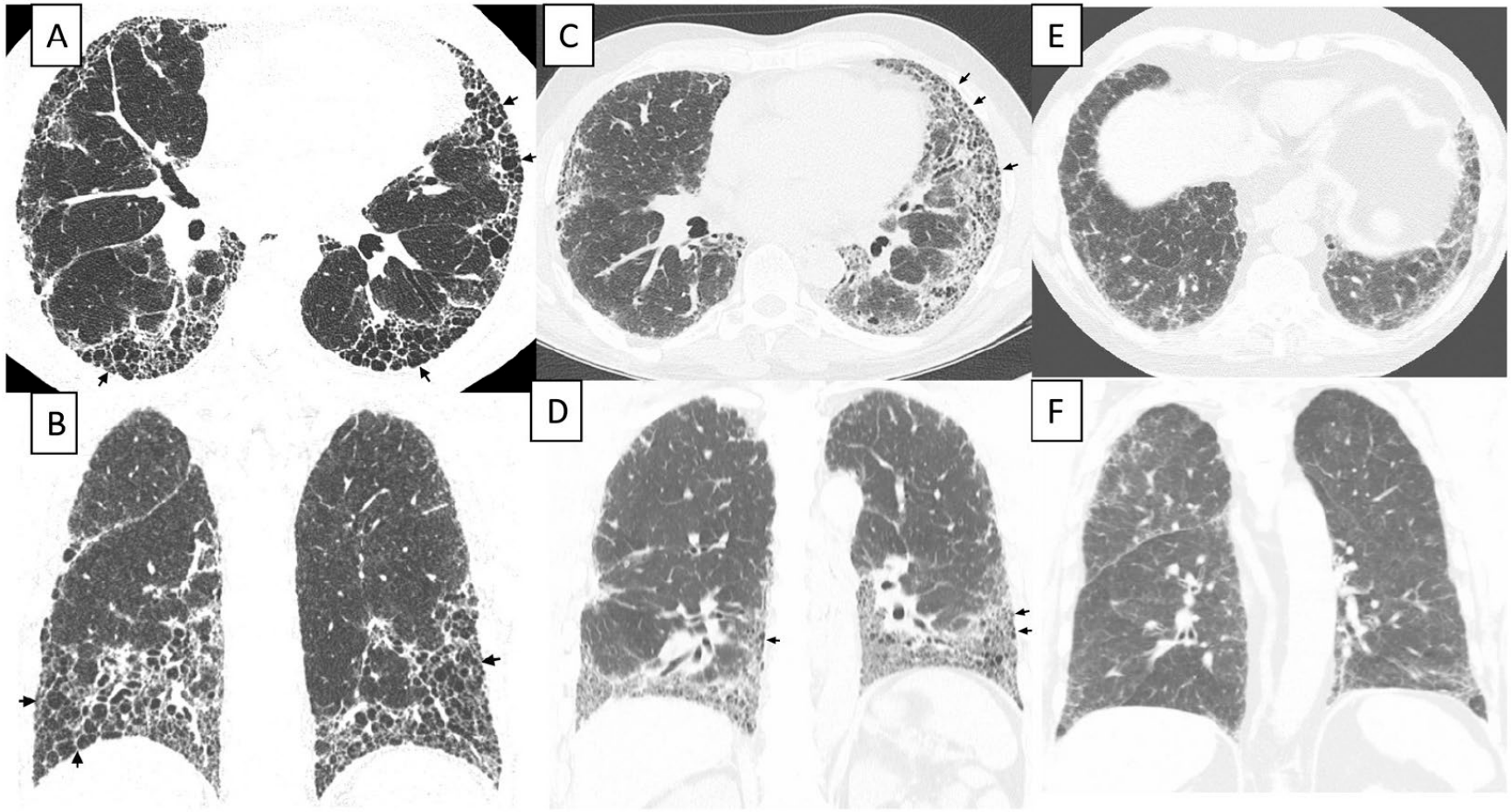
59. Vancheri C et al. *Eur Respir J*. 2010;35(3):496-504.

Chest X-Ray might show:

- The radiologist reports: *"Bilateral reticulations, with peripheral and basal predominance. Suggestive of pulmonary fibrosis"*



HRCT in IPF



7. Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788-82.

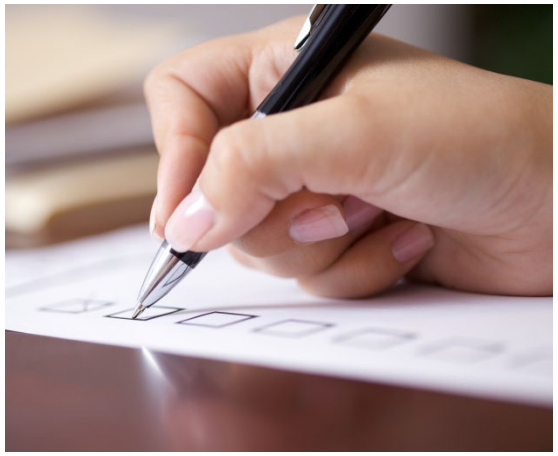
Other investigations

- **Pulmonary function testing (PFT)**
  - ↓TLC, ↓FVC (may be normal in early stages)
  - Normal-to-increased FEV1/FVC, ↓DL_{CO}
  - ↓ survival if low baseline FVC, ↓FVC, ↓DL_{CO}
- **6-minute walk test (6MWT)**
  - Measure of disease status
  - ↓6MWD → ↓survival
- **Surgical lung biopsy**
  - Rarely required (clinical picture + UIP pattern on HRCT is usually diagnostic)
  - Done if diagnosis remains uncertain despite review of clinical and radiological information

13. Oldham JM et al. *Respir Med*. 2014;108(6):819-829. 19. du Bois RM et al. *Am J Respir Crit Care Med*. 2011;183(9):1231-1237.

Delayed referral → poorer survival in IPF

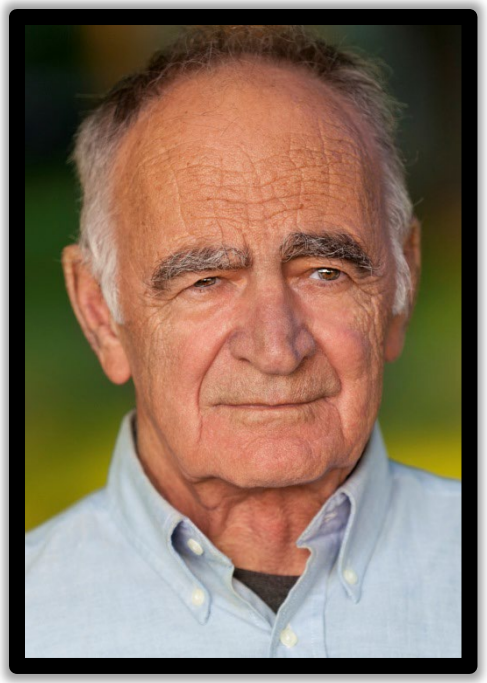
- Canadian patient survey :
  - Average wait: 20 months from symptom onset to confirmed diagnosis
  - 32% patients received 1 diagnosis other than IPF, 15% received ≥ 3 diagnoses
  - Diagnosis may be delayed ≥ 5 years
- PCPs - Key role in early diagnosis, referral to respirologist, and in some cases, ordering HRCT thorax



2. Sgalla G et al. *Respirology*. 2016;21(3):427-37. 23. http://cpff.ca. 24. Zibrak JD et al. *NPJ Prim Care Respir Med*. 2014;24:14054.

The Patient revisited

- HRCT ordered; shows UIP pattern:
  - Reticular opacities with basal and subpleural predominance, associated with honeycomb appearance
- Patient referred to respirologist
- PFTs show restrictive pattern
  - ↓ FVC, ↓ TLC, and ↓ DL_{CO}
- No cause for UIP pattern of ILD identified
- Diagnosed as IPF



Case challenge

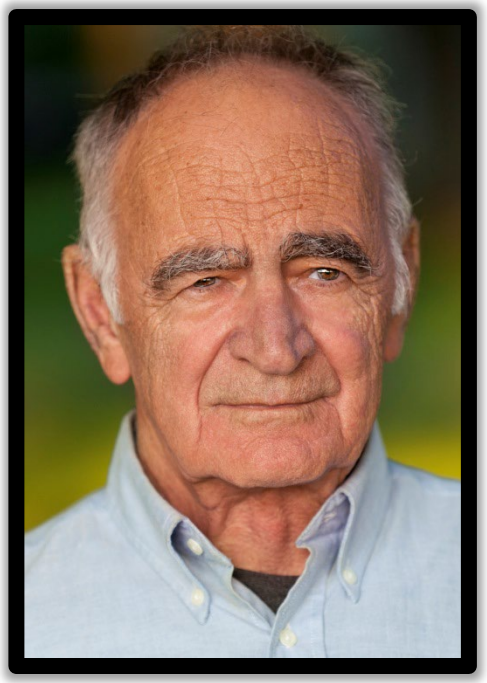
The most common clinical presentation of IPF involves:

a) Productive cough and fever

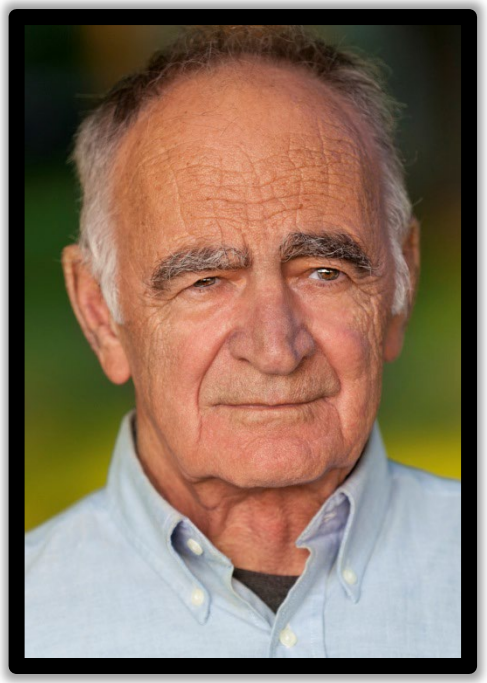
b) Cough, exertional dyspnea, inspiratory crackles, finger clubbing

c) Acute respiratory distress

d) Dyspnea at rest



Case challenge



The most common clinical presentation of IPF involves:

a) Productive cough and fever

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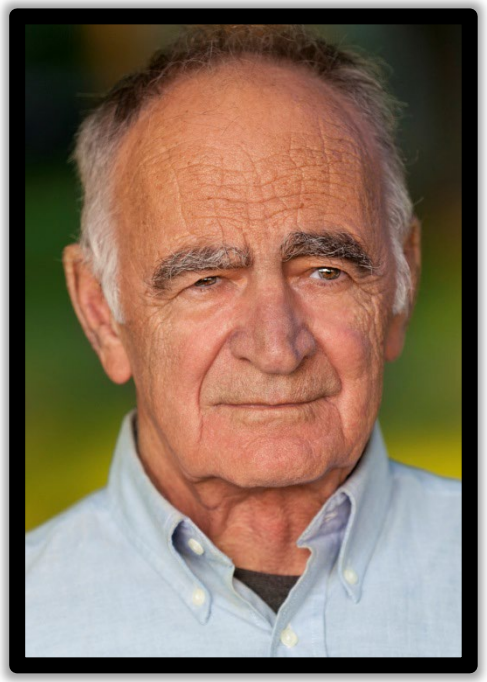
Case challenge

Approved pharmacological treatment options for IPF include:

a) Pirfenidone and nintedanib
b) Prednisolone and cyclophosphamide
c) Azathioprine and N-acetylcysteine
d) Bronchodilators



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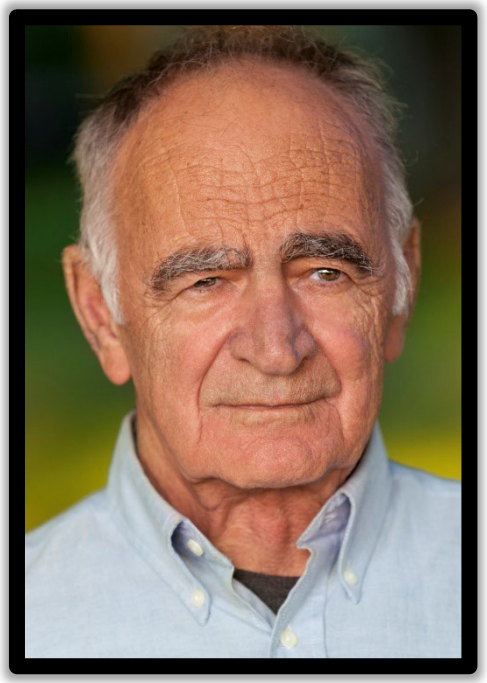


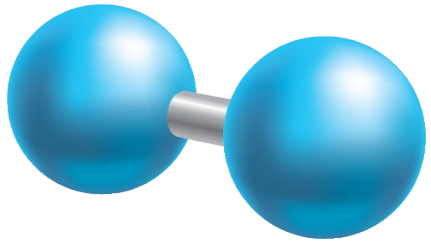
Approved pharmacologic therapies for IPF

- ✓ Pirfenidone: Antifibrotic agent with pleiotropic effects
- ✓ Nintedanib: Tyrosine Kinase Inhibitor (inhibits PDGFR, FGFR, VEGFR)

1. Raghu G et al. *Am J Respir Crit Care Med*. 2015;192(2):e3-19. 7. Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788-824. 29. Wells A. *Eur Respir J*. 2015;45(5):1208-1210.

Oxygen therapy in IPF

- Hypoxemic at rest → impaired QoL
- No controlled studies of long-term oxygen for resting hypoxemia in IPF
- Indirect evidence from studies showing survival benefit in obstructive lung disease
- If clinically significant resting hypoxemia → long-term oxygen therapy
- Some evidence for oxygen use in patients who only desaturate during exercise; improved exercise endurance



7. Raghu G et al. Am J Respir Crit Care Med. 2011;183(6):788-824.
62. Arizono S et al. Eur Respir J. 2015;46(suppl 59).

Oxygen

Oxygen desaturation contributes to decreased exercise

In patients who desaturate with exercise <88% and were administered supplemental oxygen to maintain saturation >90%

- Led to farther walk distance
- Reduced dyspnea scores and fatigue

Many patients' attitude changed about oxygen after experiencing increased ability to exercise

Decreased cough incidence

Khor, et al. *ERJ*. 2024.
Visca, et al. *Lancet Resp*. 2018

Roadblocks to oxygen usage

- Psychological burdens
- Fear of running out
- Embarrassment of being seen with oxygen in public
- Perceived disease is worsening with use of oxygen
- Unsure what types to use

Khor, et al. *ERJ*. 2024.
Visca, et al. *Lancet Resp*. 2018

Oxygen devices

Stationary concentrator

Stationary liquid concentrator

Compressed gas tank

Portable concentrator

Portable liquid concentrator

Nasal cannula tubing

Cough

Can be multifactorial

- ILD, GERD, allergic rhinitis, cough receptor sensitivity

When all other possibilities are exhausted, then can treat with neuromodulators

- Gabapentin, amitryptilline, baclofen

Other medication options

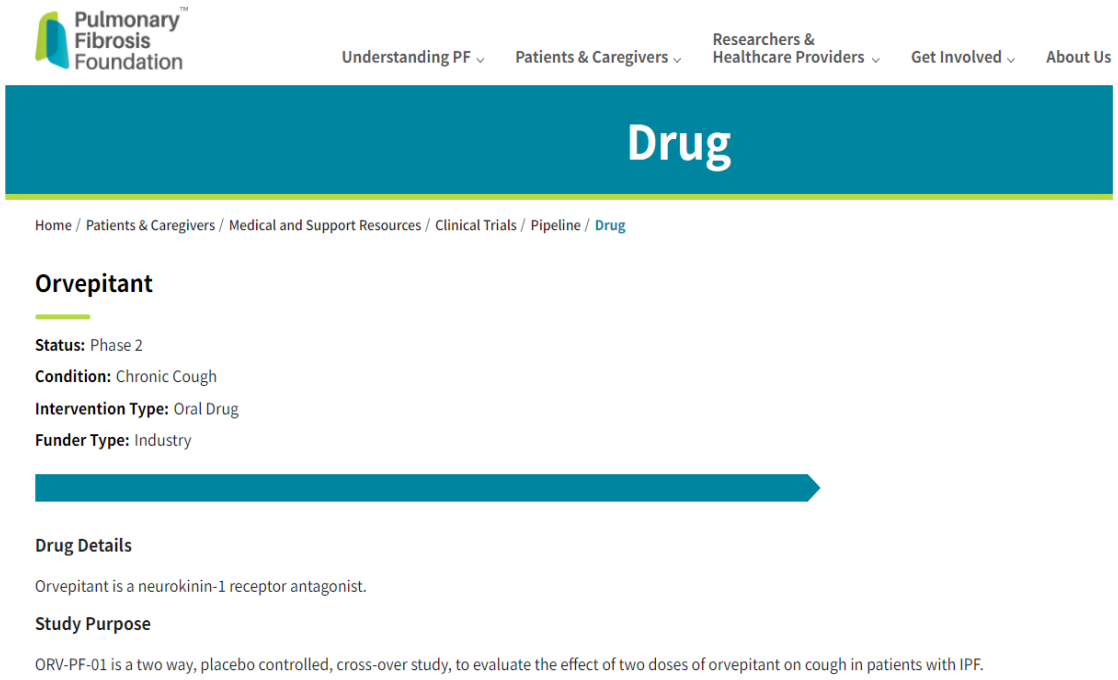
- Opioids, benzodiazepines

Clinical trials ongoing

- Orvepitant

Khor, et al. *ERJ*. 2024.

Newer neuropathic agents for cough



Pulmonary Fibrosis Foundation

Understanding PF | Patients & Caregivers | Researchers & Healthcare Providers | Get Involved | About Us

Drug

Home / Patients & Caregivers / Medical and Support Resources / Clinical Trials / Pipeline / Drug

Orvepitant

Status: Phase 2

Condition: Chronic Cough

Intervention Type: Oral Drug

Funder Type: Industry

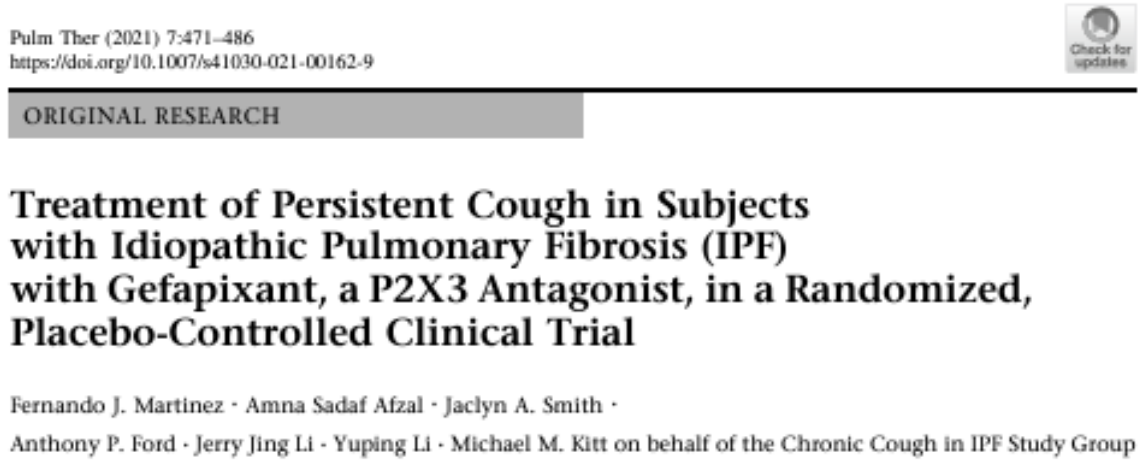
Drug Details

Orvepitant is a neurokinin-1 receptor antagonist.

Study Purpose

ORV-PF-01 is a two way, placebo controlled, cross-over study, to evaluate the effect of two doses of orvepitant on cough in patients with IPF.

https://www.pulmonaryfibrosis.org/patients-caregivers/medical-and-support-resources/clinical-trials-education-center/pipeline/drug/other-related-conditions/orvepitant



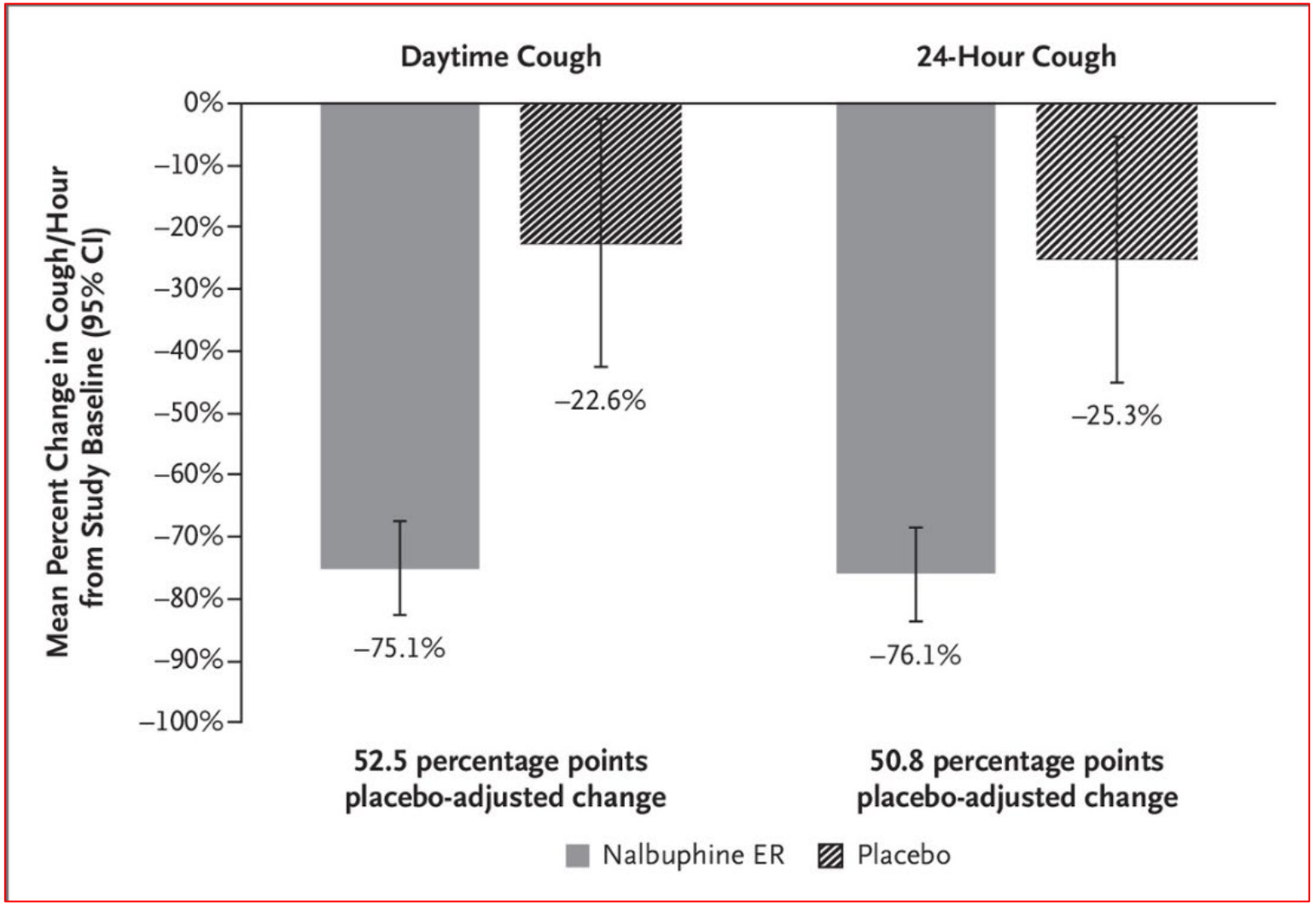
Pulm Ther (2021) 7:471–486
https://doi.org/10.1007/s41030-021-00162-9

ORIGINAL RESEARCH

Treatment of Persistent Cough in Subjects with Idiopathic Pulmonary Fibrosis (IPF) with Gefapixant, a P2X3 Antagonist, in a Randomized, Placebo-Controlled Clinical Trial

Fernando J. Martinez · Amna Sadaf Afzal · Jaclyn A. Smith · Anthony P. Ford · Jerry Jing Li · Yuping Li · Michael M. Kitt on behalf of the Chronic Cough in IPF Study Group

Martinez FJ, Afzal AS, Smith JA, Ford AP, Li JJ, Li Y, Kitt MM; Chronic Cough in IPF Study Group. Treatment of Persistent Cough in Subjects with Idiopathic Pulmonary Fibrosis (IPF) with Gefapixant, a P2X3 Antagonist, in a Randomized, Placebo-Controlled Clinical Trial. Pulm Ther. 2021 Dec;7(2):471-486. doi: 10.1007/s41030-021-00162-9. Epub 2021 Jun 21. PMID: 34152585; PMCID: PMC8589896.



NEJM Evidence

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ORIGINAL ARTICLE

Nalbuphine Tablets for Cough in Patients with Idiopathic Pulmonary Fibrosis

Authors: Toby M. Maher, M.D., Ph.D., Cristina Avram, M.D., Enoch Bortey, Ph.D., Simon P. Hart, M.D., Ph.D., Nikhil Hirani, M.D., Ph.D., Philip L. Molyneux, M.D., Ph.D., Joanna C. Porter, M.D., Ph.D., Jaclyn A. Smith, M.D., Ph.D., and Thomas Sciascia, M.D. Author Info & Affiliations

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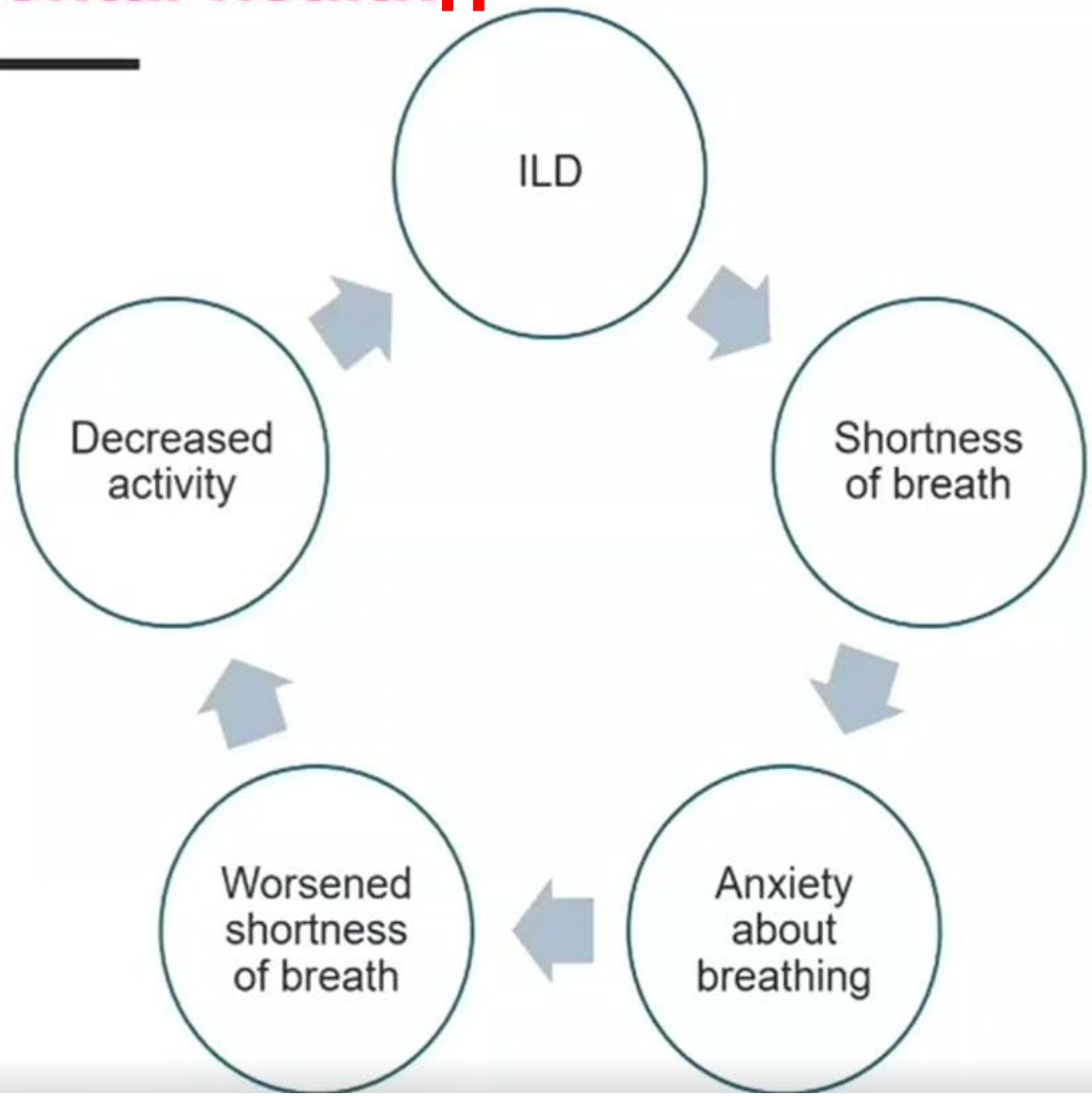
Maher TM, Avram C, Bortey E, Hart SP, Hirani N, Molyneux PL, Porter JC, Smith JA, Sciascia T. Nalbuphine Tablets for Cough in Patients with Idiopathic Pulmonary Fibrosis. NEJM Evid. 2023 Aug;2(8):EVIDoa2300083. doi: 10.1056/EVIDoa2300083. Epub 2023 May 22. PMID: 38320144.

Mental health!!



- Therapy
- Palliative care team
- Medications

Wisjenbeek, et al. *AJRCCM*. 2019.

Lung transplantation in IPF

- Improves survival
- Offered to appropriate patients
- Adult lung recipients median survival 5.5 years
- Lung Allocation Score (LAS)*
  - Based on medical urgency and post transplant survival
  - Used to prioritize candidates for transplantation
- Early referral for evaluation

*Although version of LAS is used, it has not been standardized in Canada.

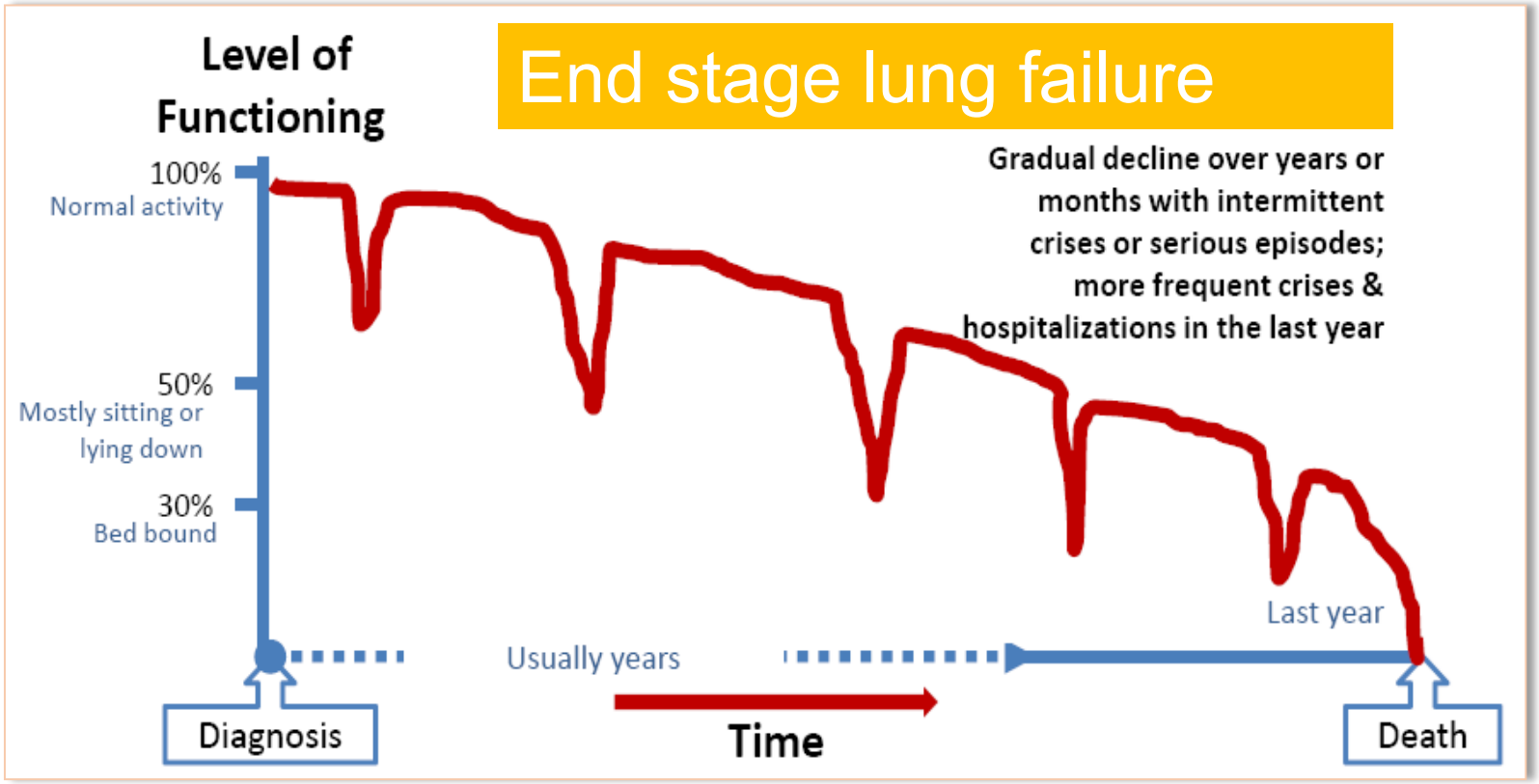
7. Raghu G et al. *Am J Respir Crit Care Med*. 2011;183(6):788-824. 46. Adamali HI et al. *Curr Respir Care Rep* 2012;1:208-215. 49. Thabut G et al. *J Thoracic Cardiovasc Surg*. 2001;126(2):469-475.

Acute exacerbations of IPF

- Acute respiratory deterioration with diffuse airspace changes on chest imaging either triggered (e.g. infection, post-op, drug) or idiopathic
- High in-hospital mortality associated with exacerbations
- Need to rule out pulmonary emboli (need CT angiogram)
- Supportive therapy is mainstay
- High dose corticosteroids commonly used, but limited data to support efficacy
- Pirfenidone, nintedanib not used to treat exacerbations
- Immunosuppressants, anticoagulants – inconclusive

7. Raghu G et al. *Am J Respir Crit Care Med.* 2011;183(6):788-824. 50. Kim D. *Respir Res.* 2013;14(1):86. 15. Collard HR, et al. *Am J Respir Crit Care Med* 2016;194:265–275. 58. Bhatti H et al. *Ann Thorac Med.* 2013;8(2):71-77.

Life trajectory



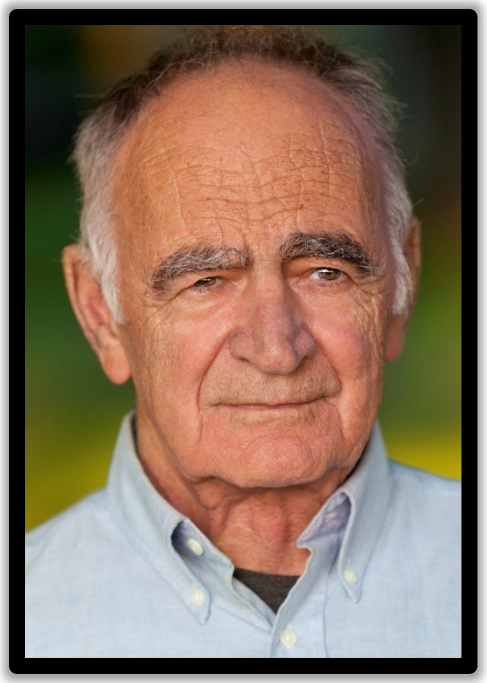
Comorbidities in IPF

- Impact on QoL, survival
- GERD: PPIs for patients with GERD symptoms
- Obstructive sleep apnea: CPAP improves QoL
- Pulmonary hypertension (PH): No benefit of treating PH with PAH specific drugs on IPF outcomes
- Emphysema: Increases mortality
- Mood disturbances: Screening, management recommended
- Lung cancer: Adversely affects survival

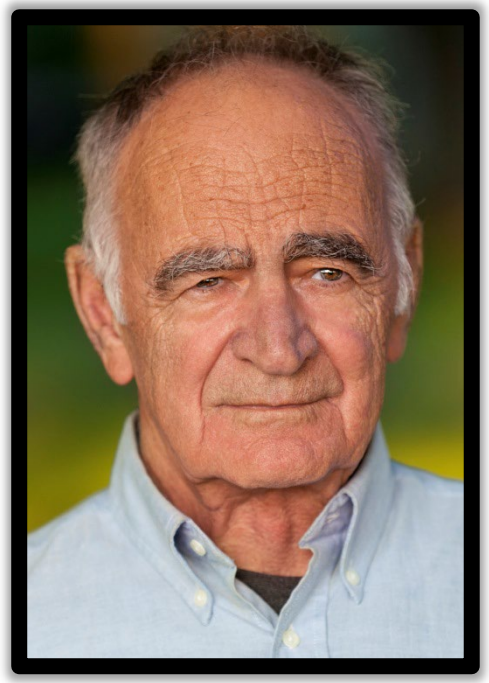
1. Raghu G et al. *Am J Respir Crit Care Med.* 2015;192(2):e3-19. 13. Oldham JM et al. *Respir Med.* 2014;108(6):819-829. 51. Lee JS et al. *Am J Respir Crit Care Med.* 2011;184(12):1390-1394. 52. Mermigkis C et al. *Sleep and Breathing.* 2013;17(4):1137-1143. 53. Zisman DA et al. *N Engl J Med.* 2010;363(7):620-628. 54. King TE et al. *Am J Respir Crit Care Med.* 2011;184(1):92-99. 55. Glaspole I et al. *A94. New Insights in IPF*; 2016:A2606-A2606. 56. Akhtar AA et al. *Chron Respir Dis.* 2013;10(3):127-133. 61. Tomassetti S et al. *Chest.* 2015;147(1):157-164. 63. Mejía M et al. *Chest.* 2009;136(1):10-15.

Revisit the patient

- Patient advised about treatment options
- Nintedanib 150 mg twice daily initiated
- Shared care: PCP monitors patient every 3 months for disease progression and complications, treatment related adverse effects
- **One month later:**
  - Patient complains of diarrhea



Case challenge



For patients experiencing diarrhea with nintedanib:

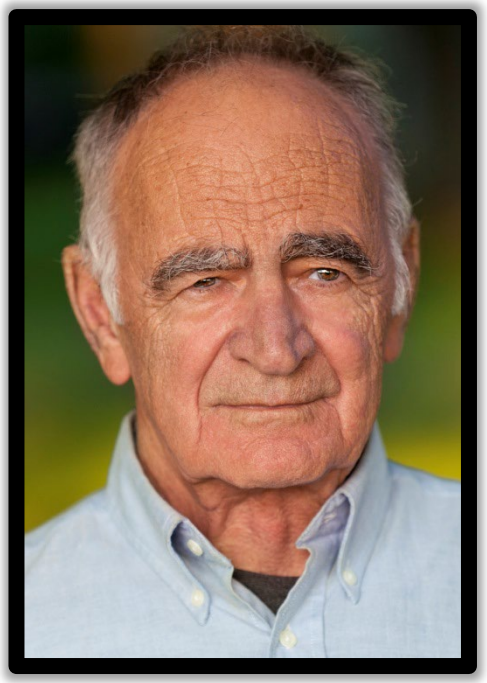
a) Discontinuation of treatment is the only option.

b) Antidiarrheal medication (e.g., loperamide) is not recommended.

c) Treatment with an opiate is recommended.

d) Consider dosage reduction, treatment interruption, or discontinuation if diarrhea does not respond to conventional antidiarrheal management.

Case challenge



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Management of key adverse effects

- Pirfenidone
  - Nausea/vomiting: Take with food
  - Photosensitivity: Avoid/minimize exposure to sunlight
- Nintedanib
  - Diarrhea: Manage at first sign of symptoms; start antidiarrheal (e.g., loperamide)
  - Nausea/vomiting: Treat (e.g., with antiemetics) as needed

Source: Prescribing information OFEV, ESBRIET

Key learning points

- Early diagnosis needed; delay in referral to specialist care associated with poorer survival
- All chronic cough is not COPD; all ILD is not IPF
- Refer (earlier!) relevant patients to respirologist for appropriate diagnosis and treatment
- Progressive disease, will need palliative care at some point!

If unexplained cough/dyspnea and suspect ILD… REFER!

Integrating Palliative Care Principles into Pulmonary Fibrosis Management

What can be your trigger to have the palliative care discussion?

- At diagnosis of lung disease
- Signs of disease progression
- At diagnosis of advanced disease
- Considering ICU
- When quality of life worsens
- Transplant being considered or planned.
- Increase symptom burden
- Expressions of frustration
- Changes in health status
- Exacerbations or progression of co-morbidities (HF, kidney disease)
- New co-morbidities (e.g. stroke, MI)
- Increase in frailty
- Falls
- Reduction in mobility
- Reduced weight
- Change in cognitive function
- Hospitalizations or recurrent ED visits.

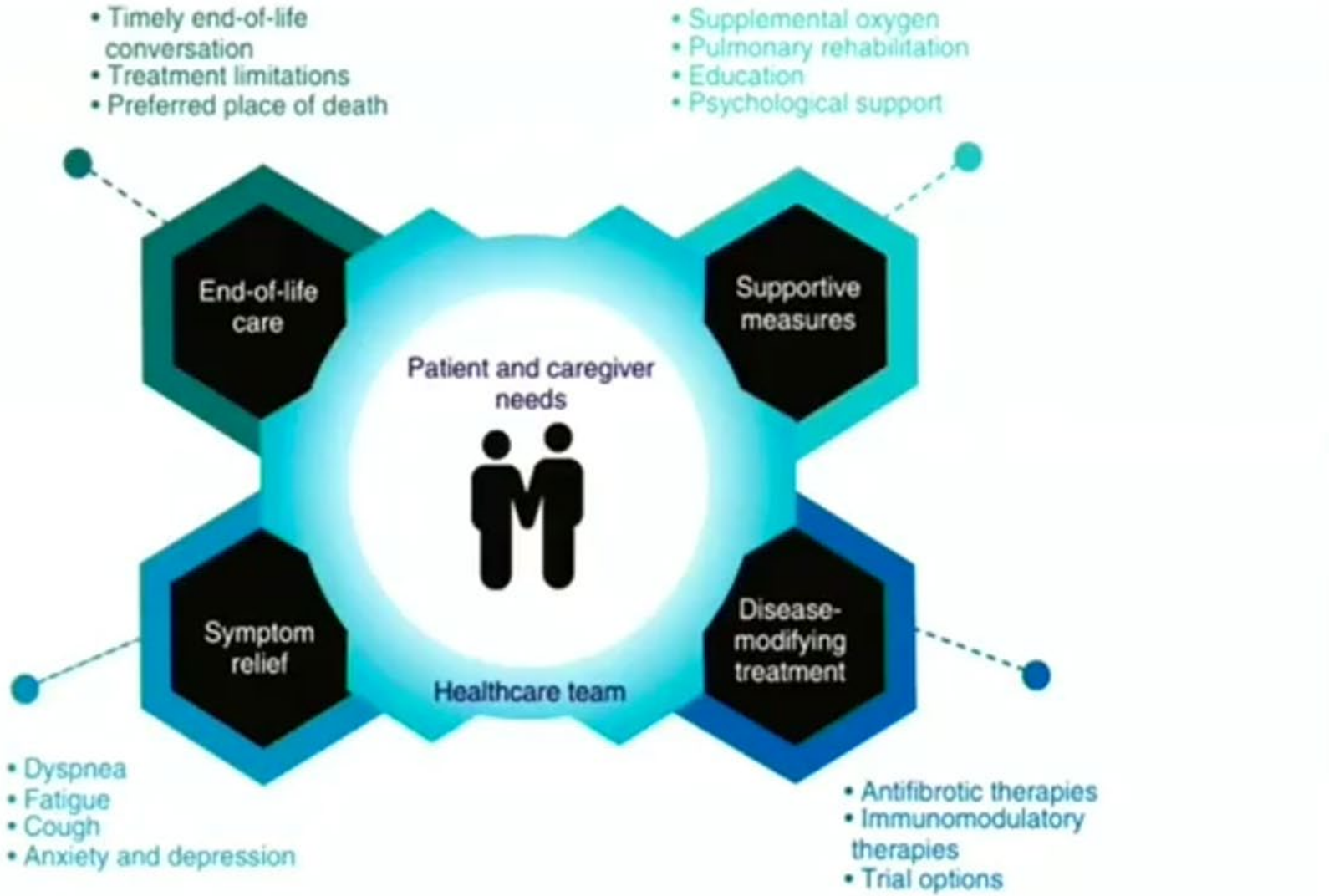
Would you be surprised if your patient died in the next year?

Palliative care in IPF

- ↓ burden of symptoms
- Consider referral if significant decline in lung function/functional status requiring assistance for ADL
- Home care teams can assist in providing symptom relief and advance care planning for end

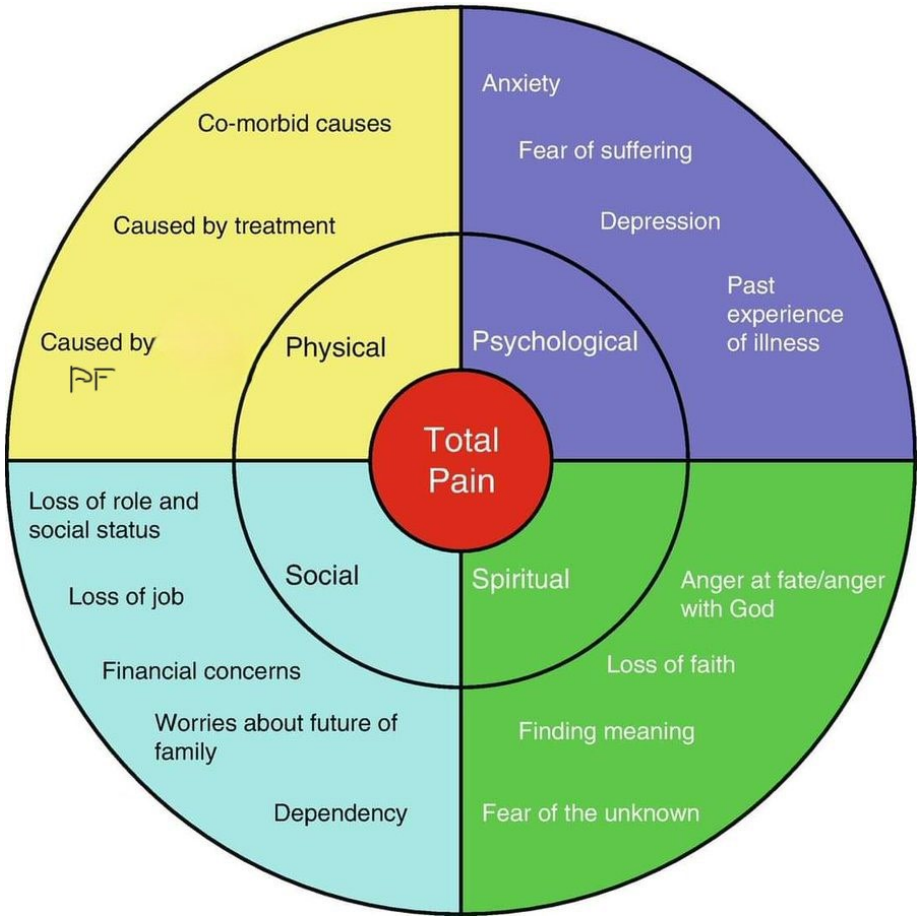
45. Thickett DR et al. *Thorax*. 2014;69(12):1136-40. 46. Adamali HI. *Curr Respir Care Rep* 2012;1:208-215

Approach



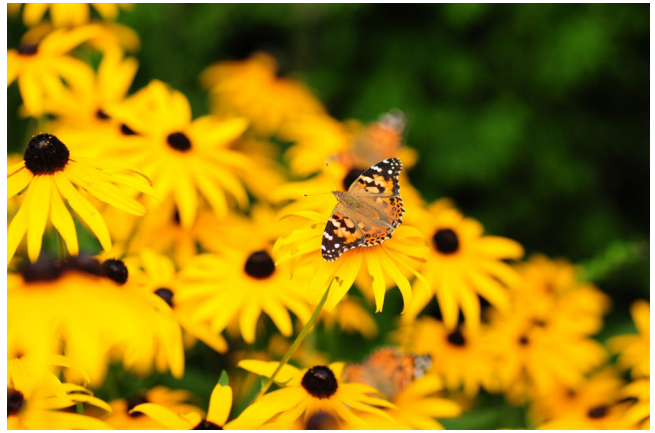
Wisjenbeek, et al. *AJRCCM*. 2019

Integrating Palliative Care Principles into Pulmonary Fibrosis Management - continued



Conversations

- Advance Care Planning:
  - Designate an attorney for personal care
  - Reflect and discuss/communicate wishes, goals, “what is important”
- Assess and Review Understanding of Illness:
  - Illness Trajectory
  - Expectations
  - Prognosis
- Goals of Care Conversations:
  - General directions for care based on wishes/goals
- Treatment-Related Decision-Making:
  - Based on GOCs and current situation/status of illness



Interdisciplinary Collaboration

Beyond Idiopathic Pulmonary Fibrosis Diagnosis: Multidisciplinary Care With an Early Integrated Palliative Approach Is Associated With a Decrease in Acute Care Utilization and Hospital Deaths

Journal of Pain and Symptom Management. 2018 Feb;55(2):420-426.

Conclusions: MDC care model for IPF was associated with reduced health care use in the last year of life and more home deaths.

INTERDISCIPLINARY COLLABORATION

- Social Worker: Finances; WSIB; adjustment
- Respiratory Therapist: Oxygen equipment; health teaching
- Physiotherapist: Energy-conservation; chest physio; maintenance
- Occupational Therapist: Energy-conservation
- Nursing: Smoking cessation; Symptom mngt; monitoring; ACP/GOC
- Medicine

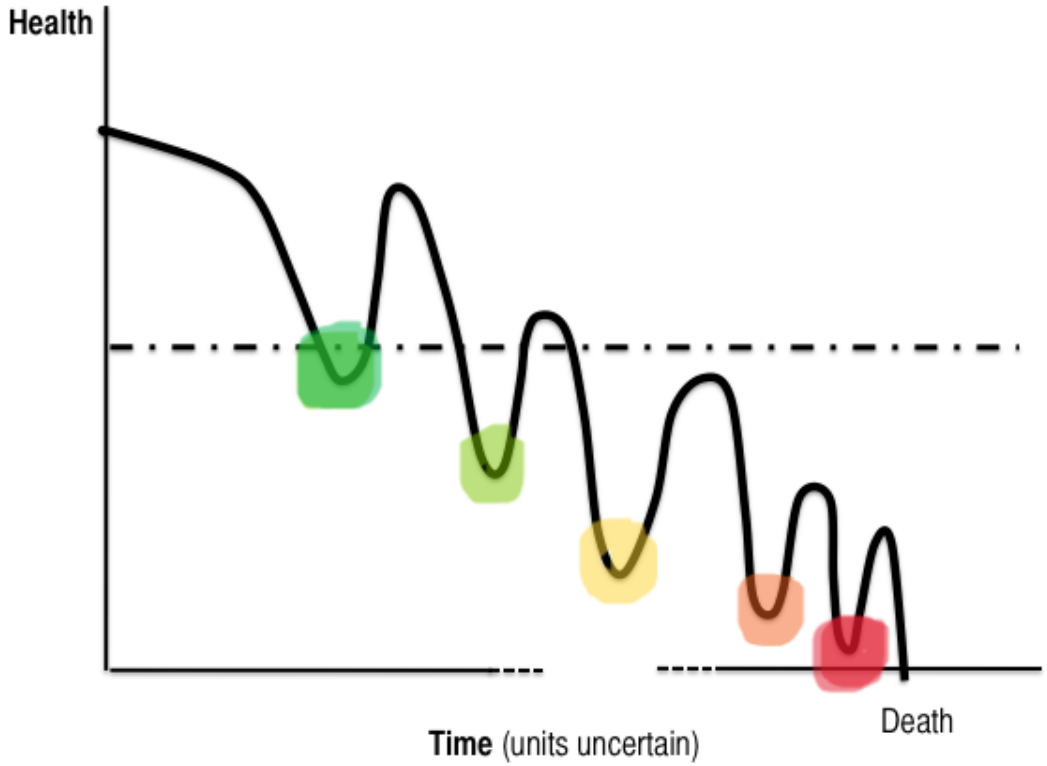
Pulmonary Rehabilitation

- Exercise
- Review of COPD Action Plan
- Understand Breathing Techniques
- Oxygen and Spirometry
- Managing Stress and Anxiety
- How to Conserve Energy (planning, pacing, prioritizing)
- Nutrition and Lifestyle
- Impact on Family and Relationships
- Review of Medications
- Strategies to Improve Sexual Activity
- Introduction of Advanced Care Planning and Palliative Care

Symptom Management tips and tricks (including approach at EOL)

- Sudden death from sudden respiratory or cardiac event/acute exacerbation

- Death from disease progression and respiratory failure



Common Symptoms with Pulmonary Fibrosis

- Hypoxemia*
- Dyspnea
- Anxiety
- Cough/Respiratory Secretions

Oxygen – tips and tricks

- Hypoxemia ≠ dyspnea (to a certain extent)
- Assessing hypoxemia (rest, **exertion**, overnight)
- Can tolerate high levels of 02*** (difficulties in increasing oxygen before exertion and reducing it)
- At home: can go up to 14-15L/min by combining 2 concentrators of up to 10L/min each, usually 7 and 7)
- Can add humidity to concentrator (usually if higher than 6L/min)
- Involve RT for proper delivery equipment (right nasal prongs/masks/equipment)
- Outside mobility very limited (short life and burden of portable tanks and canisters) and anxiety-provoking
- Can be connected to CPAP
- EoL: Stop monitoring oxygen

Dyspnea – tips and trick

- Assessment:
  *Hypoxemia ≠ dyspnea
  *Increased WOB ≠ dyspnea: SUBJECTIVE SYMPTOM
  *Descriptors
  - Severity
  - Onset/Timing/duration
  - Triggering/alleviating
  - Concurrent symptoms
  - Impact

- Manage underlying cause(s) and contributing cause(s)

- Non-pharm measures:
  - fan/blowing air/ventilation/circulating air
  - positioning (especially at night)
  - pacing and pausing, limiting activities*
  - Meditation/relaxation*

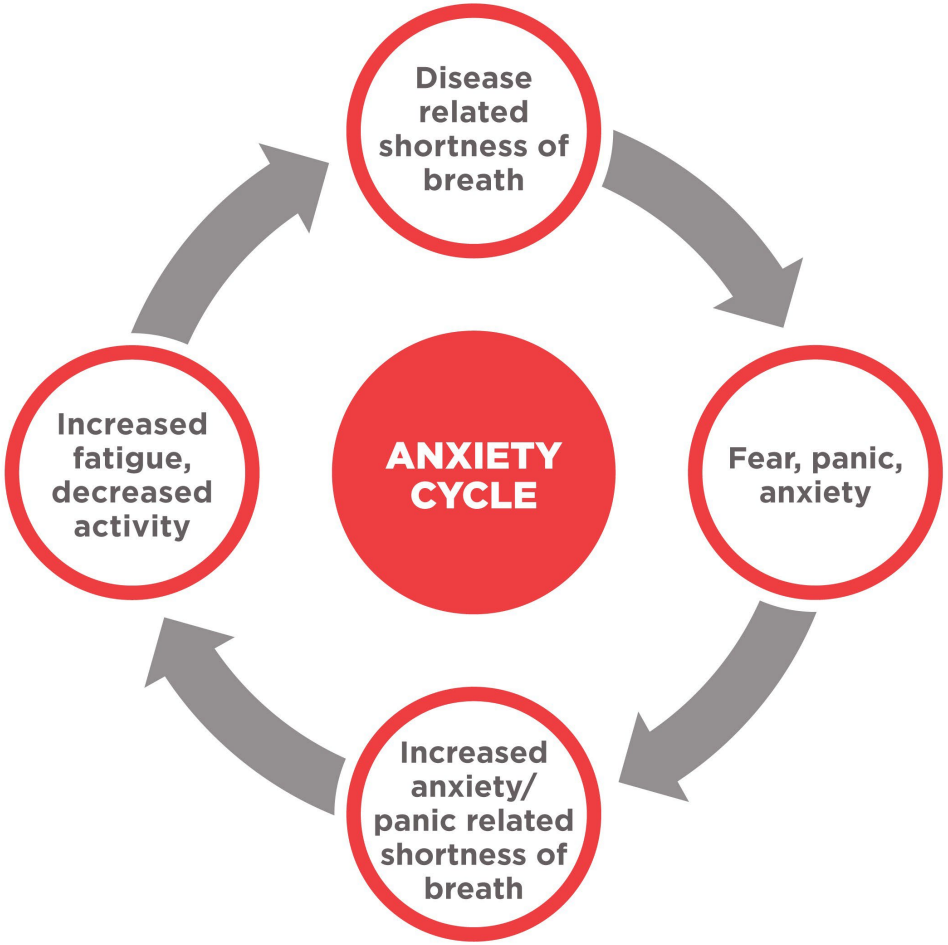
Dyspnea – tips and trick - Continued

Pharm:

- Oxygen
- Opioids:
  - Start low and slow
  - Lower doses are usually required for dyspnea than for analgesia, but in our experience not always true in PF
  - Often need for rapid escalation (depending on disease progression)
  - Consider regularly scheduled or CR formulation
  - Order PRN for breakthrough dyspnea and pre-exertion
  - Consider CADD for constant dyspnea, severe exertional dyspnea, with advanced disease and at EOL
- ?methotrimeprazine (sedative/anxiolytic effect Vs direct effect on dyspnea?)

Dyspnea-related anxiety – tips and tricks

- Very prominent in PF, more severe than in COPD (in our experience)
- Manage dyspnea first
- Low dose benzodiazepines (clonazepam, lorazepam) during the day if tolerated for constant/easily-triggered anxiety
- SL/SC midazolam for exertional anxiety
- CADD at EOL or earlier (?rebound anxiety)
- Social presence/reassurance



Cough and secretions– tips and tricks

- Assessment:
  - Dry/irritated/non-productive cough Vs difficult to clear secretions/productive?
  - Frequency, severity, triggers, impact, etc…
  - Untreated underlying cause? (E.g. post-nasal drip, GERD, dysphagia, asthma, smoking, infection, medications etc…)

- Dry/irritated cough:
  - All opioids will act as cough suppressants (lower doses usually needed than for analgesia)
  - Experiential/anecdotal evidence suggesting elixir opioids can work better (?soothing ?faster absorption)
  - Saline nebs; humidified oxygen; humidifiers*
  - Consider antitussives

Cough and secretions– tips and tricks - Continued

- **Tenacious secretions (not common):**
  - Humidity: Saline nebs; humidified oxygen; humidifiers*
  - Mucolytics:
    - Acetylcysteine* 3–10 mL of 10-20% solution nebulized four times daily
    - Guaifenesin
  - Lung Clearance strategies
  - Deep breathing/coughing exercises
  - Vibrating devices

Mrs M.

- Woman in her early 60s
- Current smoker, although reduced
- Co-morbid COPD
- Dxed 5 years ago with ILD, did not tolerate pirfenidone and nintedanib, not a candidate for lung transplant due to smoking and other co-morbs.
- PMHx: PTSD/anxiety, GERD, chronic pain
- Social hx: lives alone, 2 adult children but limited support

Mrs. M.

- TIMELINES:

  - JANUARY: Consultation to community palliative care for dyspnea
    1. Continue low dose hydromorphone (HM) for chronic pain for dyspnea
    2. Introduced PRN HM doses for breakthrough dyspnea
    3. Review non-pharm strategies for dyspnea
    4. Continue PRN oxygen 2L/min during exertion
    5. Mirtazapine at HS for anxiety/sleep (pre-existing + disease-related)

  - FEBRUARY:
    1. Started on hydromorphone Contin (HM CR) for worsening dyspnea
    2. Worsening dyspnea-related anxiety
    3. Home PT and OT for ADL equipment, strengthening, energy-conservation

  - BEGINNING OF MARCH:
    1. HM CR dose increased for worsening dyspnea
    2. Consideration of SL midazolam for worsening anxiety
    3. Wears oxygen almost constantly at 2L/min

Mrs. M.

- TIMELINES:

  - END OF MARCH:
    1. Worsening dyspnea on exertion
    2. SC HM injections started for faster onset
    3. PSW support put in place for ADLs
    4. SL midazolam started for worsening anxiety
    5. Saline nebs for new cough

  - BEGINNING OF APRIL:
    1. SL midazolam switched to SC midazolam (pt's preference)
    2. SW involvement re: anxiety, living situation, finances

  - MID-APRIL:
    1. CADD HM and midazolam started for worsening dyspnea and anxiety
    2. Oxygen requirements increasing, now at 4.5L/min constant

Mrs. M.

- TIMELINES:

  - END OF APRIL TO END OF JUNE:
    1. Oxygen requirements increasing, RT in to R/A, 10L concentrator brought in
    2. CADD HM and midazolam up-titration
    3. Increasing care needs at home (meals, hygiene, toileting)
    4. Trials of living with family, did not work out

  JULY: Moved to hospice

  What happened in hospice?

Questions?

Wrap Up

- Please fill out the feedback survey following the session! Link has been added into the chat.
- A recording of this session will be e-mailed to registrants within the next week.
- Please join us for the next session in this series on **Symptom management in advanced respiratory illnesses held on September 18th, 2024, from 12–1:00 p.m. ET.**

Thank You

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