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IPF: What Do We Know and What Do We Need to Know?

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A Symposium for Those Living with IPF
ILD Collaborative
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4 Burning Questions in Idiopathic Pulmonary Fibrosis

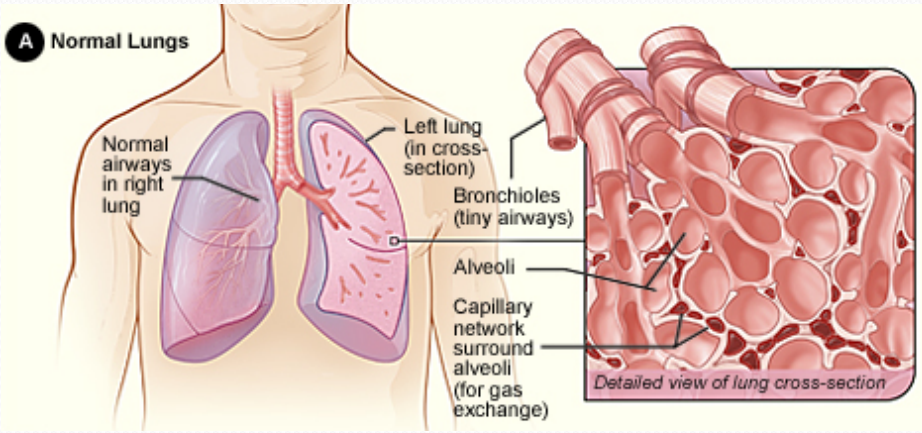
- What is IPF?
- How did I get it?
- What is going to happen to me?
- What can we do about it?

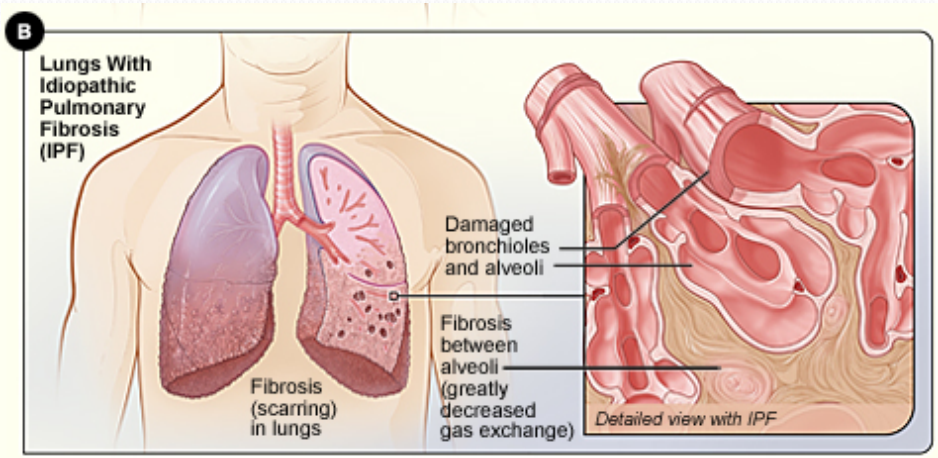
What is IPF?

Idiopathic (unknown cause)

Pulmonary (lungs)

Fibrosis (scarring)





Images from: National Heart, Lung and Blood Institute

What is IPF?

"Diagnosis of exclusion"

Requires exclusion of known (i.e. non-idiopathic) causes of pulmonary fibrosis:

- Infections
- Exposures (e.g. asbestos, radiation, medications)
- Autoimmune diseases (e.g. rheumatoid arthritis)

Can look (and behave) just like IPF!

How is IPF Diagnosed?

- Clinical History (symptoms, exposures)
- Physical Exam
- Laboratory testing

Evaluating for other causes of pulmonary fibrosis

- Chest CT scan – one of 3 categories
  - Definite IPF → lung biopsy often not needed
  - Possible IPF → lung biopsy may help determine IPF vs. other
  - Not IPF

- Pulmonary function tests (PFTs) – assess disease severity and monitor for change over time

How Did I Get IPF?

Short Answer: We don't know….

…but we have some ideas.

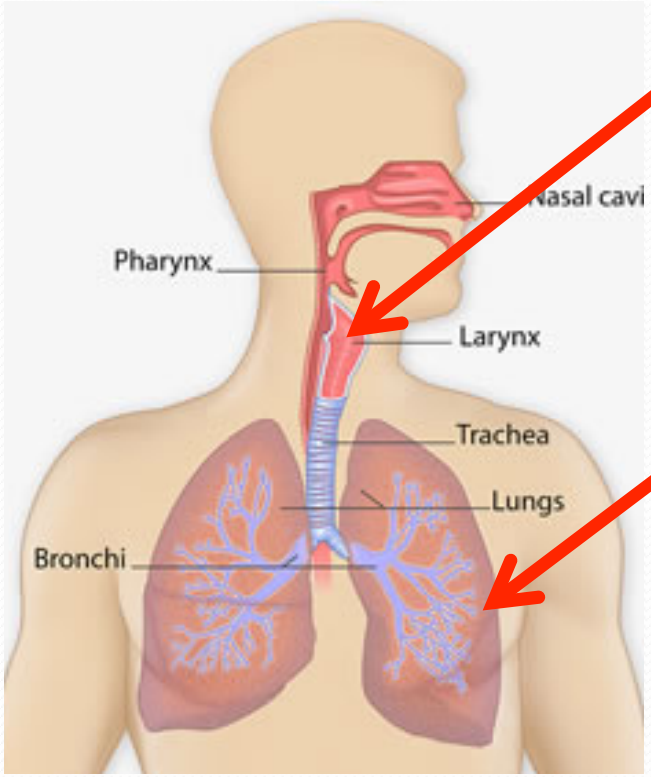


Image from: National Heart, Lung and Blood Institute

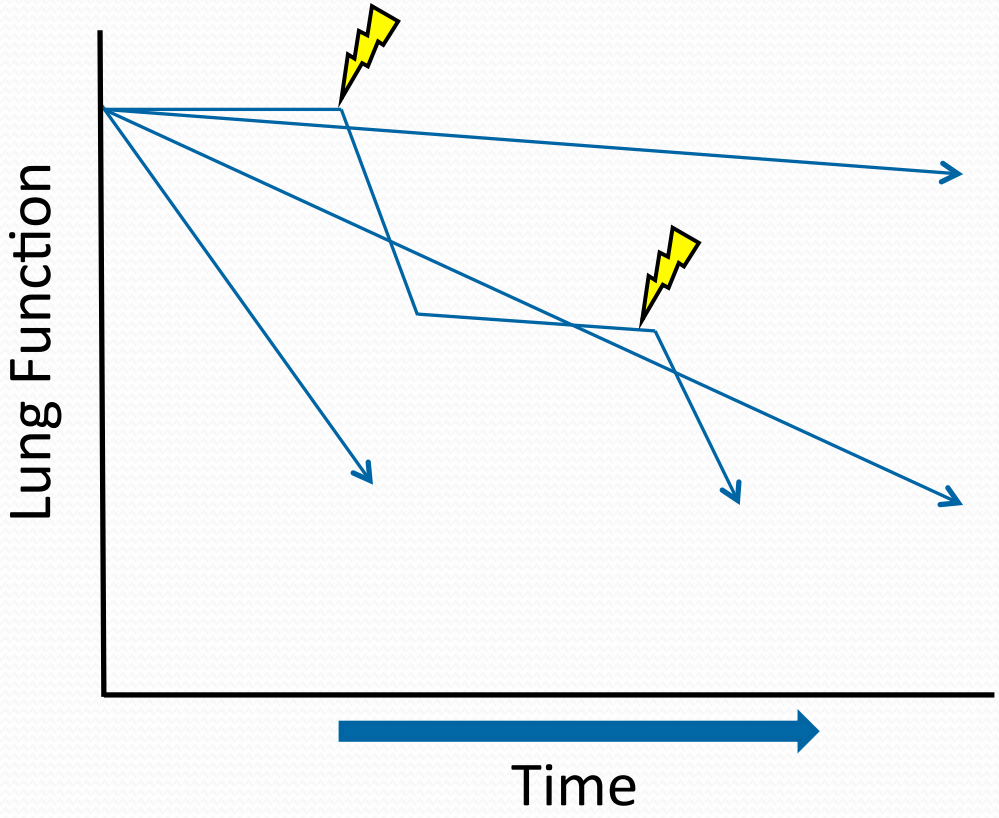
Ongoing, repetitive lung injury
- Stomach acid (acid reflux)?
- Environmental exposures?
- Viruses?

Excessive scar formation
- "Overactive wound-healing"?
- Genetic predisposition?
- Accelerated aging in the lung?

What is Going to Happen to Me?

Accumulation of more scarring in the lungs over time…

….but *pace* of progression is highly variable



Key questions:

1. How can we predict who is going to experience rapid vs. slow progression of disease?
2. How can we predict acute exacerbations?
3. Will current (and future) therapies preferentially benefit one group vs. another?

Adapted from Ley et al. *AJRCCM.* 2011.

What can we do about it?

- New anti-fibrotic therapies (Drs. Zibrak and LaCamera)
- Treating associated illnesses (Dr. Montesi)
- Investigational drugs (Dr. Tager)
- Relaxation Techniques (Ms. Malloy)
- Symptom management, oxygen use, and exercise/pulmonary rehab (small groups)
- Lung transplantation (Dr. Astor)