10:30 – 11:45 am

Idiopathic Pulmonary Fibrosis: Raising the Index of Suspicion in Primary Care

SPEAKER

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Presenter Disclosure Information

The following relationships exist related to this presentation:

► Gary M. Hunninghake, MD: Medical Advisory Board for "Patient's Like Me." Consultant for Medna, LLC, George Lehman Group.

Off-Label/Investigational Discussion

► In accordance with pmiCME policy, faculty have been asked to disclose discussion of unlabeled or unapproved use(s) of drugs or devices during the course of their presentations.

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Objectives

- Describe the epidemiology and pathophysiology of idiopathic pulmonary fibrosis (IPF)
- Recognize the signs and symptoms of IPF and identify patients in whom referral and further evaluation for IPF may be warranted
- Outline current diagnostic criteria and the importance of early identification of IPF
- Summarize data for recently approved pharmacologic agents for the treatment of IPF

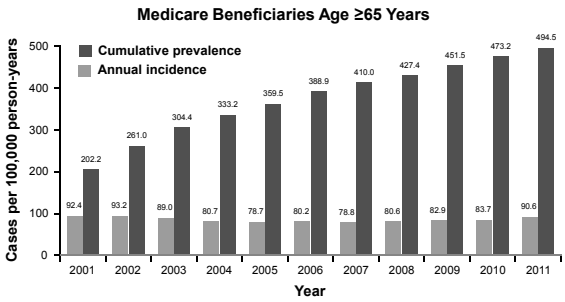
Epidemiology of Idiopathic Pulmonary Fibrosis (IPF)

Epidemiology of IPF

- Approximately 5 million patients worldwide[1]
- Numbers in the U.S.[2]
  - Incidence: >50,000 patient/year
  - Prevalence: >130,000 patients currently
- More common in men than women[1,2]
- Mean age at presentation is 66 years[1,2]
  - Two-thirds of all cases diagnosed in patients >60 yrs old
- Median survival[1,2]
  - 2–5 years

1. Meltzer EB, Noble PW. *Orphanet J Rare Dis*. 2008;3:8. 2. Raghu G, et al. *Lancet Respir Med*. 2014;2:566–572.

Increasing Prevalence of IPF



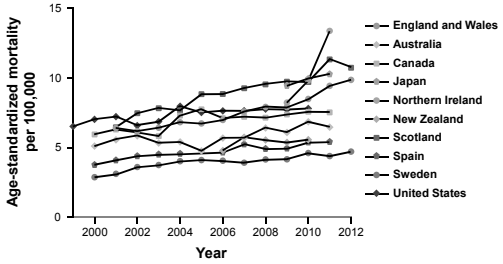
Factors associated with lower survival

- Age, index year, male gender

Median survival = 3.8 years

Raghu G, et al. *Lancet Respir Med*. 2014;2:566–572.

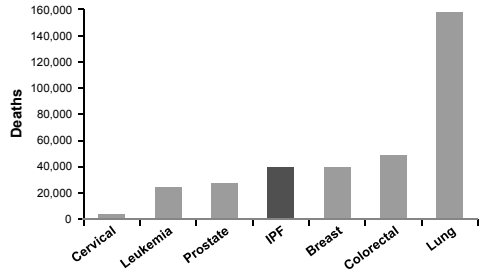
Idiopathic Pulmonary Fibrosis
Increasing Global Mortality



- Age-standardized mortality ~4 and 10 per 100,000 population
- Overall 2–3% annual increase in mortality
- 28,000–65,000 deaths in Europe, and 13,000–17,000 deaths in the U.S. from IPF in 2014

Hutchinson JP, et al. *Ann Am Thorac Soc*. 2014;11(8):1176–1185.

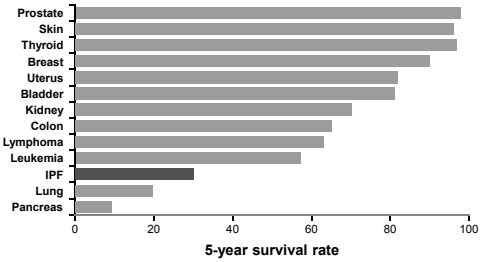
Estimated Deaths:
IPF[1] vs Common Cancers[2]



1. Coalition for Pulmonary Fibrosis. Facts About Idiopathic Pulmonary Fibrosis. Available at: http://www.coalitionforpf.org/facts-about-idiopathic-pulmonary-fibrosis/. 2. American Cancer Society, Surveillance and Health Services Research, 2015. Available at: http://www.cancer.org/acs/groups/content/@editorial/documents/document/acspc-044552.pdf.

5-Year Mortality Rate High in IPF

- 5-year survival rate (U.S.) is only 20–40%[1]
- Worse survival rates than many common cancers



1. Reprinted with permission from Vancheri, et al. *Eur Respir J*. 2010; 35(3):496-504.

Signs and Symptoms of IPF

What Is IPF?

- Idiopathic pulmonary fibrosis (IPF) is a chronic lung disease that belongs to a family of lung disorders known as interstitial lung diseases (ILD), and within this family, IPF is part of a subgroup known as the idiopathic interstitial pneumonias (IIP).
- IPF is characterized by scar tissue within the lungs of unknown cause and is associated with a pathologic pattern of usual interstitial pneumonia (UIP)

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824. Meltzer EB, Noble PW. *Orphanet J Rare Dis*. 2008;3:8.

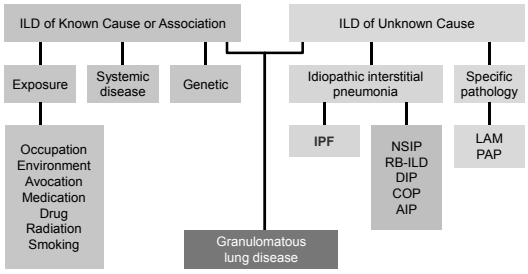
Interstitial Lung Disease (ILD)

- Diverse group of disorders that involve the pulmonary parenchyma; also known as diffuse parenchymal lung disease (DPLD)
- Typical presentation
  - Progressive dyspnea and dry cough
  - Abnormal pulmonary physiology
  - Abnormal CXR and/or HRCT
- Etiology
  - Idiopathic
  - Systemic diseases (connective tissue disorders)
  - Toxic, radiologic, environmental, occupational exposures

CXR = chest x-ray; HRCT = high-resolution computed tomography.

http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/pulmonary/interstitial-lung-disease/Default.htm. July 8, 2015.

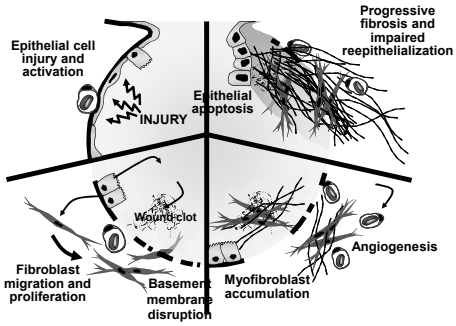
Classification of ILD



IIP = idiopathic interstitial pneumonia; NSIP = nonspecific IP; RB-ILD = respiratory bronchiolitis ILD; DIP = desquamative IP; COP = cryptogenic organizing pneumonia; AIP = acute IP; LAM = lymphangioleiomyomatosis; PAP = pulmonary alveolar proteinosis.

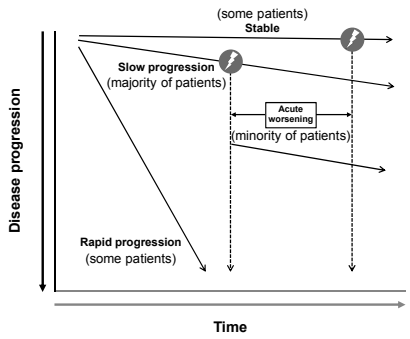
http://www.clevelandclinicmeded.com/medicalpubs/diseasemanagement/pulmonary/interstitial-lung-disease/Default.htm. July 8, 2015. Travis WD, et al. *Am J Respir Crit Care Med*. 2013;188(6):733–748.

Pathogenesis of IPF



Ahluwalia N, et al. *Am J Respir Crit Care Med*. 2014;190(8):867–78.

Natural History of IPF



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

Common Risk Factors for IPF

- Age
- Male gender
- Cigarette smoking
- Gastroesophageal reflux
- Occupational exposure to metal dust or wood dust
- ?Viral infection
- Genetic predisposition

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824. http://www.coalitionforpf.org/epidemiology-and-risk-factors/. Accessed July 6, 2015.

How Do IPF Patients Present?

- Shortness of breath (dyspnea)
- Dry cough
- Fatigue
- "Velcro" rales at lung bases
- Clubbing of fingers and/or toes may be present
- Incidentally
  - ILD on routine CXR or CT chest
  - ILD at bases of abdominal CT
  - Fluoroscopy at time of cardiac catheterization
  - Family history

CXR = chest x-ray; CT = computed tomography.

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824. Meltzer EB, Noble PW. *Orphanet J Rare Dis*. 2008;3:8. http://my.clevelandclinic.org/health/diseases_conditions/hic-idiopathic-pulmonary-fibrosis. Accessed July 6, 2015.

When to Suspect IPF?

- Susceptible individual:
  - Family history of ILD
  - Exposures
- Symptoms:
  - Shortness of breath
  - Cough — usually dry and can be intractable
- Physical examination:
  - Incidental inspiratory crackles on exam ("Velcro"-like)
- Pulmonary function tests: Restrictive lung disease on PFTs
- CXR or CT consistent with "chronic interstitial changes"
- Exercise desaturation

PFT = pulmonary function test.

Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824. Meltzer EB, Noble PW. *Orphanet J Rare Dis*. 2008;3:8. http://www.nhlbi.nih.gov/health/health-topics/topics/ipf. Accessed July 6, 2015.

Diagnosis of IPF

Dyspnea Differential

Upper airways
- Laryngospasm
- Vocal cord dysfunction ⬅
- Airway obstruction
- Tracheomalacia

Lower airways/lungs
- Asthma ⬅
- COPD ⬅
- Pneumonia ⬅
- Pneumothorax ⬅
- Pleural effusion ⬅
- Parenchymal and endobronchial tumors
- ILD ⬅
- ARDS

Cardiovascular system
- CHF ⬅
- Pulmonary edema ⬅
- Pericardial effusion
- Arrhythmias ⬅
- ↓Cardiac output
- Cardiomyopathy ⬅
- Ischemic heart dx ⬅
- Valvular disease ⬅
- Pulmonary embolism ⬅
- Pulmonary hypertension ⬅

Neuromuscular system
- Cerebrovascular events
- Amyotrophic lateral sclerosis
- Spinal cord injuries above C3
- Multiple sclerosis
- Myasthenia gravis
- Guillain-Barré syndrome
- Polymyositis
- Muscular dystrophy

Metabolic, renal, and heme
- Acidosis
- Renal failure/tubular acidosis
- Hyperthyroidism
- Hypothyroidism
- Cushing's syndrome
- Obesity
- Anemia and methemoglobin
- ARDS

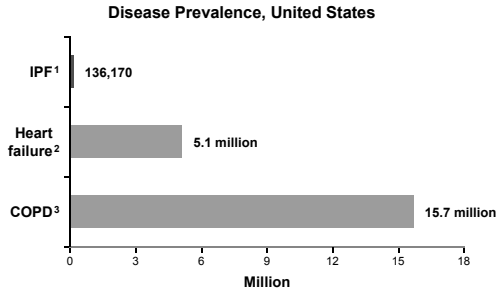
Toxins/poisons
- Botulism
- Tetanus
- Organophosphate poisoning
- CO poisoning
- Drugs

Psychogenic disorders ⬅
- Hyperventilation syndrome
- Anxiety/panic disorders
- Depression
- Somatization disorder

⬅ **= The "Biggies"**

⬅ **= The "Not so biggies, but not to be forgotten"**

Distinguishing Dyspnea: IPF Prevalence



1. Raghu G, et al. *Resp Crit Care Med*. 2006;174:810–816. 2. Go AS, et al. *Circulation*. 2013;127:e6–e245. 3. Wheaton AG, et al. *MMWR Morb Mortal Wkly Rep*. 2015;64:289–295.

Workup: Methodical Approach

- Good history essential
  - Family history of ILD
  - Smoking history
  - Exposure history
    - Organic particles: birds, hot tubs
    - Inorganic particles: occupational (e.g asbestos, silica)
- CXR is a good screen
- **HRCT is essential**
- Serologic evaluation
- Surgical lung biopsy if needed

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

Pulmonary Function Tests

- Spirometry
  - Reduced forced vital capacity (FVC) and total lung capacity (TLC)
  - Normal or increased FEV_1/FVC ratio
- Impaired gas exchange
  - Decreased DL_{CO}, PaO_2
  - Desaturation on exercise oximetry
  - Increased A-aPO_2 gradient
- Normal PFTs do not exclude ILD
  - May have high baseline lung function
  - Element of emphysema in one-third of IPF patients
    - "pseudonormalization" of lung function

FEV = forced expiratory volume; DL_{CO} = diffusing capacity of the lung for carbon monoxide; PaO_2 = arterial oxygen partial pressure; A-aPO_2 = alveolar–arterial oxygen difference.

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

2011 ATS/ERS Diagnostic Criteria for IPF

Exclusion of known causes of ILD

AND

UIP pattern on HRCT without surgical biopsy

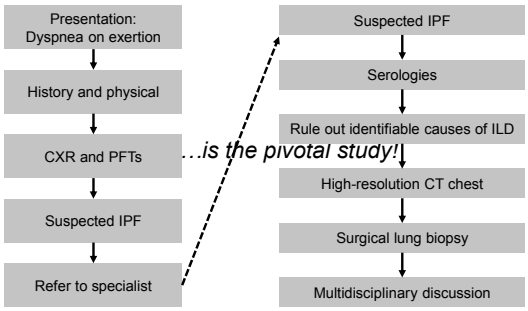
OR

Definite/possible UIP pattern on HRCT with a surgical lung biopsy showing definite/probable UIP*

***Multidisciplinary discussion among ILD experts is recommended because it increases accuracy of IPF diagnosis.**
UIP = usual interstitial pneumonia; HRCT = high-resolution computed tomography.

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

Diagnostic Algorithm for IPF



PFT = pulmonary function test.

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

HRCT Criteria for UIP Pattern

UIP Pattern
(all four features)

- Subpleural, basal predominance
- Reticular abnormality
- Honeycombing with or without traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern

HRCT = high-resolution computed tomography; UIP = usual interstitial pneumonia.

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

HRCT Criteria for UIP Pattern

Possible UIP Pattern
(all three features)

- Subpleural, basal predominance
- Reticular abnormality
- Absence of features listed as inconsistent with UIP pattern

HRCT = high-resolution computed tomography; UIP = usual interstitial pneumonia.

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

HRCT Criteria for UIP Pattern

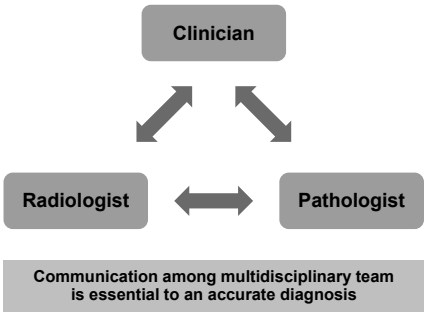
Inconsistent with UIP Pattern
(any of the seven features)

- Upper or mid-lung predominance
- Peribronchovascular predominance
- Extensive ground glass abnormality (extent >reticular abnormality)
- Profuse micronodules (bilateral, predominantly upper lobes)
- Discrete cysts (multiple, bilateral, away from areas of honeycombing)
- Diffuse mosaic attenuation/air-trapping (bilateral, in three or more lobes)
- Consolidation in bronchopulmonary segment(s)/lobe(s)

HRCT = high-resolution computed tomography; UIP = usual interstitial pneumonia.

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

IPF: An Interdisciplinary Diagnosis



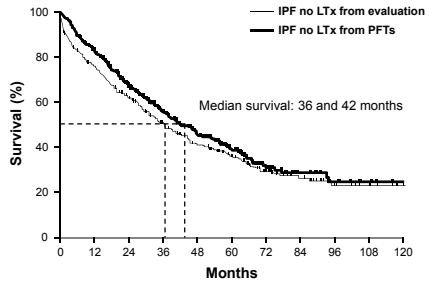
Communication among multidisciplinary team is essential to an accurate diagnosis

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

Early Diagnosis Matters in IPF

Imre Noth, MD
Professor of Medicine
University of Chicago
Chicago, IL

IPF: Survival in the Current Era
2000–2009 (n = 521)



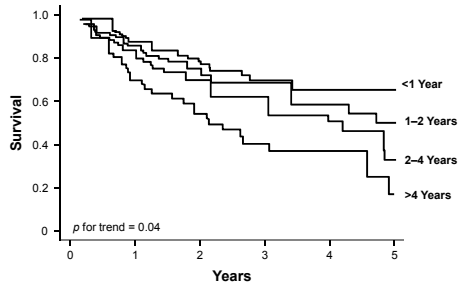
Nathan SD, et al. *Chest*. 2011;140(1):221–229.

Why Refer Early to a Specialist or Specialized Center?

- Diagnostic expertise
  - Standardized assessment
  - Confirmation of diagnosis
- Management expertise
  - Choice of an appropriate therapy
  - Oxygen prescription
  - Pulmonary rehabilitation
  - Attention to obesity and sarcopenia/frailty
  - Potential enrollment in a clinical trial
  - Transplant evaluation

Flaherty, et al. *Am J Respir Crit Care Med*. 2004;170:904–910. Flaherty, et al. *Am J Respir Crit Care Med*. 2007; 175:1054–1060. Lamas, et al. *Am J Respir Crit Care Med*. 2011;184:842–847.

Higher Mortality Associated With Delays in Accessing Care



Lamas DJ, et al. *Am J Respir Crit Care Med*. 2011;184:842–847.

Comorbid Conditions Common in IPF

- Pulmonary hypertension
- Gastroesophageal reflux disease
- Coronary artery disease
- Emphysema
- Obstructive sleep apnea

It is unknown if treating these comorbidities improves outcomes

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

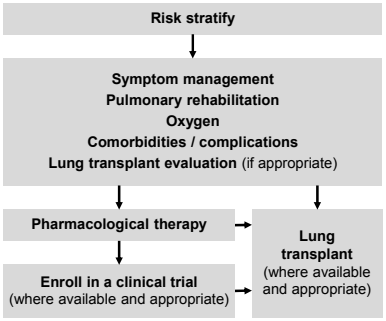
Treatment of IPF

Goals of Treatment

- Stabilize or reduce the rate of disease progression
- Reduce symptoms
- Recognize and manage acute exacerbations
- Prompt referral to lung transplant centers in select patients
- Manage comorbidities

Richeldi L. *Eur Respir Rev*. 2013;22(128):103–105. Raghu G, et al. *Am J Respir Crit Care Med*. 2011;183(6):788–824.

IPF Management Algorithm



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

2015 Guidelines
Recommendations for Management of IPF

Agent	2015 Guideline	2011 Guideline
New and Revised Recommendations		
Anticoagulation (warfarin)	Strong recommendation against use*	Conditional recommendation against use‡
Combination prednisone + azathioprine + *N*-acetylcysteine	Strong recommendation against use†	Conditional recommendation against use†
Selective endothelin receptor antagonist (ambrisentan)	Strong recommendation against use†	Not addressed
Imatinib, a tyrosine kinase inhibitor with one target	Strong recommendation against use*	Not addressed
Nintedanib, a tyrosine kinase inhibitor with multiple targets	Conditional recommendation for use*	Not addressed
Pirfenidone	Conditional recommendation for use*	Conditional recommendation against use†
Dual endothelin receptor antagonists (macitentan, bosentan)	Conditional recommendation against use†	Strong recommendation against use*
Phosphodiesterase-5 inhibitor (sildenafil)	Conditional recommendation against use*	Not addressed

***Moderate confidence in effect estimates. †Low confidence in effect estimates. ‡Very low confidence in effect estimates.**

Raghu G, et al. *Am J Respir Crit Care Med*. 2015;192(2):e3–e19.

2015 Guidelines
Recommendations for Management of IPF

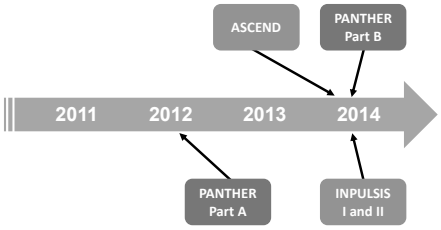
Agent	2015 Guideline	2011 Guideline
Unchanged Recommendations		
Antacid therapy	Conditional recommendation for use‡	Conditional recommendation for use‡
N-acetylcysteine monotherapy	Conditional recommendation against use†	Conditional recommendation against use†
Antipulmonary hypertension therapy for idiopathic pulmonary fibrosis-associated pulmonary hypertension	Reassessment of the previous Recommendation was deferred	Conditional recommendation against use‡
Lung transplantation: single vs bilateral lung transplantation	Formulation of a recommendation for single vs bilateral lung transplantation was deferred	Not addressed

***Moderate confidence in effect estimates. †Low confidence in effect estimates. ‡Very low confidence in effect estimates.**

Raghu G, et al. *Am J Respir Crit Care Med*. 2015;192(2):e3–e19.

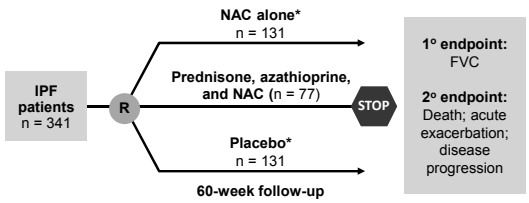
Results of Key IPF Trials

Major Phase 3 IPF Trials



PANTHER Study

IPF patients ages 35–85 years with FVC ≥ 50%, DL_{co} ≥ 3% were randomized to either *N*-acetylcysteine (NAC) alone; prednisone, azathioprine, and NAC combination; or placebo. Combination therapy stopped early.

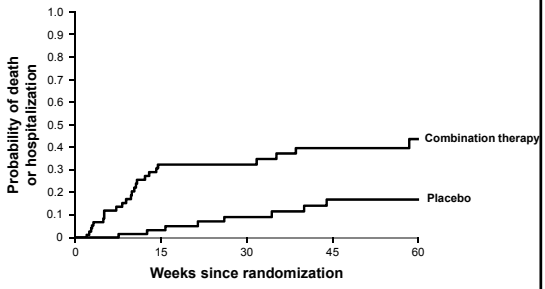


***At time of clinical alert, NAC alone (n = 81) and placebo (n = 78).**
PANTHER = Prednisone, Azathioprine and *N*-acetylcysteine: A Study That Evaluates Response in IPF; FVC = forced vital capacity.

Raghu G, et al. *N Engl J Med*. 2012;366(21):1968–1977. Martinez FJ, et al. *N Engl J Med*. 2014;370(22):2093–2101.

PANTHER Part A: Results

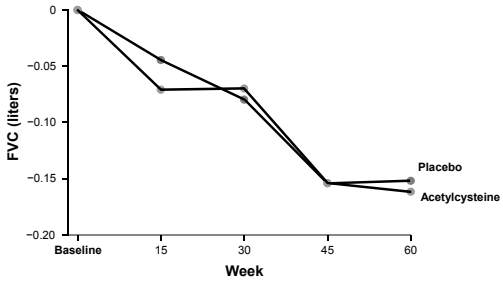
Combination therapy of prednisone, azathioprine, and NAC was stopped early for evidence of harm



Raghu G, et al. *N Engl J Med*. 2012;366(21):1968–1977.

PANTHER Part B: Results

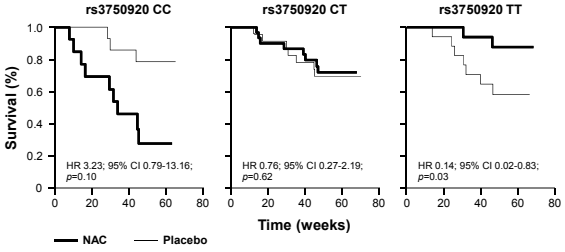
NAC alone showed no difference in FVC decline or any secondary endpoints compared to placebo



Martinez FJ, et al. *N Engl J Med*. 2014;370(22):2093–2101.

NAC Effectiveness by TOLLIP Genotype

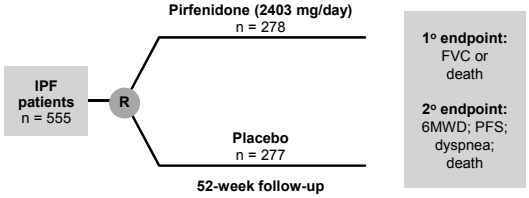
Composite endpoint of FVC decline, hospitalization, death or transplant



Oldham JM, Ma SF, Martinez FJ, Anstrom KJ, Raghu G, Schwartz DA, Valenzi E, Witt L, Lee C, Vij R, Huang Y, Strek ME, Noth I. *Am J Respir Crit Care Med*. 2015 Sep 2. [Epub ahead of print].

ASCEND Study of Pirfenidone

IPF patients ages 40–80 years with FVC between 50% to 90%, DL_{co} between 30% to 90% were randomized to pirfenidone or placebo, 127 sites in 9 countries



ASCEND = Assessment of Pirfenidone to Confirm Efficacy and Safety in IPF; PFS = progression free survival; 6MWD = 6-minute walk distance.

King TE, et al. *N Engl J Med*. 2014;370:2083–2092.

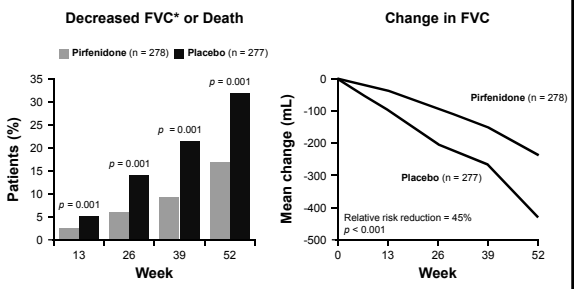
ASCEND Study: Baseline Characteristics

	Pirfenidone n = 278	Placebo n = 277
Age (years)	68	68
Male sex	80%	77%
Former smoker	66%	61%
Predicted FVC	68%	69%
FEV_1:FVC	0.84	0.84
Predicted DL_{co}	44%	44%
Dyspnea score	34	37
6MW distance (m)	415	421

FEV_1 = forced expiratory volume in one second; FVC = forced vital capacity.

King TE, et al. *N Engl J Med*. 2014;370:2083–2092.

ASCEND Study of Pirfenidone: Primary Results



***Decreased FVC is defined as an absolute decline of ≥10%.**

King TE, et al. *N Engl J Med*. 2014;370:2083–2092.

ASCEND and CAPACITY Trials of Pirfenidone: Pooled Mortality Data*

	Pirfenidone n = 623	Placebo n = 624	Hazard Ratio	*p* Value
Deaths – no. (%)				
From any cause	22 (3.5)	42 (6.7)	0.52	0.01
From respiratory cause	7 (1.1)	22 (3.5)	0.32	0.006

***Data from the two CAPACITY studies were censored at 1 year to standardize the follow-up for the three studies.**

King TE, et al. *N Engl J Med*. 2014;370:2083–2092.

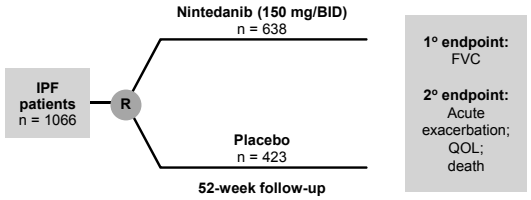
ASCEND Study: Safety and Tolerability of Pirfenidone

Adverse Event	Pirfenidone n = 623	Placebo n = 624
Nausea	36%	13.4%
Rash	28.1%	8.7%
Dizziness	17.6%	13.0%
Dyspepsia	17.6%	6.1%
Anorexia	15.8%	6.5%
Vomiting	12.9%	8.7%
Decrease in weight	12.6%	7.9%
Gastroesophageal reflux	11.9%	6.5%
Insomnia	11.2%	6.5%
Discontinuations	14.4%	10.8%
ALT and/or AST ≥3x ULN	2.9%	0.7%

King TE, et al. *N Engl J Med*. 2014;370:2083–2092.

INPULSIS 1 and 2 Studies

IPF patients ages ≥40 years with FVC ≥50%, DL_{CO} between 30% to 79% were randomized to nintedanib or placebo, 205 sites in 24 countries



BID = twice daily; QOL = quality of life.

Richeldi L, et al. *N Engl J Med*. 2014;370:2071–2082.

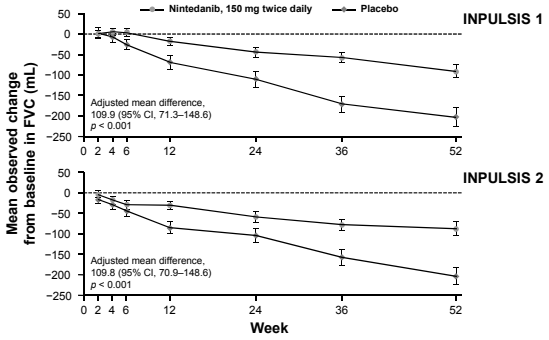
INPULSIS 1 and 2 Studies: Baseline Characteristics

	INPULSIS 1		INPULSIS 2	
	Nintedanib n = 309	Placebo n = 204	Nintedanib n = 329	Placebo n = 219
Age (years)	67	67	66	67
Male sex	81%	80%	78%	78%
Never smoked	23%	25%	31%	32%
Predicted FVC	80%	81%	80%	78%
FEV_1:FVC	0.82	0.81	0.82	0.82
Predicted DL_{CO}	48%	48%	47%	46%
Total SGRQ score*	40	40	40	39

***St. George's Respiratory Questionnaire (SGRQ) score ranges from 0 to 100, with higher scores indicating worse quality of life.**

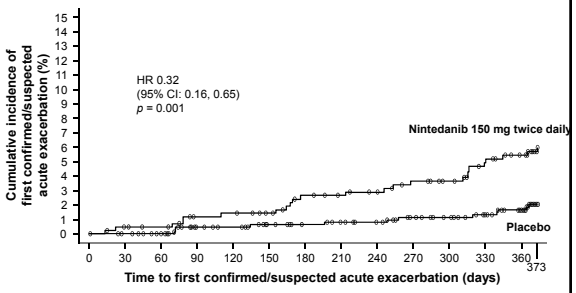
Richeldi L, et al. *N Engl J Med*. 2014;370:2071–2082.

INPULSIS 1 and 2 Studies of Nintedanib: Primary Results



Richeldi L, et al. *N Engl J Med*. 2014;370:2071–2082.

INPULSIS 1 and 2 Studies of Nintedanib:
Pooled Data on Acute Exacerbations



Richeldi L, et al. *N Engl J Med*. 2014;370:2071–2082.

INPULSIS 1 and 2 Studies:
Safety and Tolerability of Nintedanib

	INPULSIS 1		INPULSIS 2	
	Nintedanib n = 309	**Placebo** n = 204	**Nintedanib** n = 329	**Placebo** n = 219
Diarrhea	62%	19%	63%	18%
Nausea	23%	6%	26%	7%
Dyspnea	7%	11%	8%	11%
Vomiting	13%	2%	10%	3%
Weight loss	8%	6%	11%	1%
SAE	31%	27%	30%	33%
Discontinuations	21%	11%	18%	15%
ALT and/or AST ≥3x ULN	4.9%	0.5%	5.2%	0.9%

SAE = serious adverse event.

Richeldi L, et al. *N Engl J Med*. 2014;370:2071–2082.

Pirfenidone and Nintedanib

Liver function monitoring required for both pirfenidone and nintedanib

- FDA approved on October 2014 for treatment of IPF
  - No FDA approved therapies until 2014
- Pirfenidone (267 mg/cap)
  - **Dosing – Days 1–7:** 1 cap tid; **Days 8–14:** 2 caps tid; **Day 15 onward:** 3 caps tid; Maint/max: 2403 mg/day (9 caps/day)
  - **Warnings/precautions:** Photosensitivity reaction, rash, GI events, increased LFTs; pregnancy category C
- Nintedanib (100 mg and 150 mg caps)
  - **Dosing:** 150 mg bid approximately 12 hrs apart; **Max:** 300 mg/day
  - **Warnings/precautions:** GI disorders, arterial thromboembolic events, bleeding events, GI perforation, increased LFTs; pregnancy category D

http://www.pdr.net/drug-summary/esbriet?druglabelid=3632. Accessed August 3, 2015. http://www.pdr.net/drug-summary/ofev?druglabelid=3633. Accessed August 3, 2015.

IPF Management in 2015

IPF: Implications of Having Approved Drugs

- The stakes are raised!
- Need to make an accurate diagnosis
  - Avoid starting patients inappropriately on drug
- Earlier diagnosis becomes essential
  - Increased awareness
- Primary care providers: Low threshold for a pulmonary referral
- Pulmonologists: Low threshold for referral to ILD center

Managing Patient Expectations

- Discussion about what to expect from the drugs
- Efficacy
  - "The drug slows the rate of loss of lung function"
  - "You won't necessarily feel better!"
- Side effects
  - "You won't necessarily have side effects, but here is what they are."
  - **Liver function test monitoring**
    - **Pirfenidone and nintedanib**

IPF Management Checklist

- Risk factor reduction
- Patient education
  - Advocacy group involvement
- Focus on comorbidities
  - Mental health needs
  - GERD, OSA, CAD, PH, VTE, etc.
- Supplemental oxygen
- Age-appropriate vaccinations
- Discussion about available medical therapies
- Pulmonary rehabilitation
- Clinical trials
- Lung transplant evaluation
- Address end-of-life issues: palliative and hospice care

GERD = gastroesophageal reflux disease; OSA = obstructive sleep apnea; CAD = coronary artery disease; PH = pulmonary hypertension; VTE = venous thromboembolic disease.

IPF Therapies

- Pharmacologic therapy
  - Nintedanib
  - Pirfenidone
  - No head-to-head studies available for these two drugs
- Oxygen therapy
- Pulmonary rehabilitation
- Lung transplantation

Monitoring for Disease Progression

- Every 3 to 6 months by pulmonologist:
  - PFTs
  - 6-minute walk test (distance/nadir saturation)
  - O_2 requirement
  - Comorbidities
  - Consider dyspnea questionnaire (e.g., UCSD SOB questionnaire)
- HRCT for suspicion of clinical worsening

O_2 = oxygen; SOB = shortness of breath; HRCT = high-resolution computed tomography.

Take-Home Messages

- **IPF is not uncommon**
  - Not all shortness of breath is heart failure, COPD, or asthma.
- **Familiarity with signs and symptoms**
  - Recalcitrant dry cough, SOB, coarse inspiratory basilar crackles
- **Awareness and management of associated comorbidities**
  - CAD, GERD, OSA, and PH
- **There are now FDA approved novel therapies for IPF**
  - Nintedanib and pirfenidone
- **IPF is the most common indication for lung transplantation**
  - Small minority of IPF patients are candidates
- **Non-pharmacologic interventions are important**
  - Pulmonary rehabilitation and oxygen
  - Support groups
  - End-of-life planning