Idiopathic Pulmonary Fibrosis
Diagnosis & Treatment

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Disclosure Information

• I have the following financial relationships to disclose:
  Consultant for: Boehringer Ingelheim, Roche/Genentech, Veracyte, Biogen, Gilead, Pharmakea, Aeolus
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  Stockholder in: None
  Honoraria from: None
  Employee of: None

• I will not discuss off label use or investigational use in my presentation.
Outline & Objectives

• Understand the classification and diagnosis of Interstitial Lung Diseases and IPF
• Recognize typical patterns of disease on HRCT
• Discuss the potential benefits and adverse reactions of approved therapies for IPF
Interstitial Lung Diseases - Difficulties

- Diverse group of disorders (130+)
- Similar symptoms, physiology, radiology
- Difficult nomenclature
- Limited, often toxic, treatments
Distinguishing Dyspnea: IPF Prevalence

COPD: chronic obstructive pulmonary disease; IPF: idiopathic pulmonary fibrosis.
Diffuse Parenchymal Lung Disease (DPLD)

DPLD of known cause, eg, drugs or association, eg, collagen vascular disease

Idiopathic Interstitial Pneumonias

Granulomatous DPLD, eg, sarcoidosis

Other forms of DPLD, eg, LAM, HX, etc

Major

Rare

Unclassifiable

Idiopathic Pulmonary Fibrosis

Nonspecific interstitial pneumonia (idiopathic)

Acute Interstitial Pneumonia

Respiratory bronchiolitis interstitial lung disease

Desquamative Interstitial Pneumonia

Cryptogenic Organizing Pneumonia

Lymphoid Interstitial Pneumonia

Pleuroparenchymal Fibroelastosis

Diagnosis Matters!

IPF/UIP confers a poor prognosis

<table>
<thead>
<tr>
<th>Parameter</th>
<th>HR (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td>IPF diagnosis</td>
<td>28.46 (5.5, 147)</td>
</tr>
<tr>
<td>Age</td>
<td>0.99 (0.95, 1.03)</td>
</tr>
<tr>
<td>Female sex</td>
<td>0.31 (0.13, 0.72)</td>
</tr>
<tr>
<td>Smoker</td>
<td>0.30 (0.13, 0.72)</td>
</tr>
<tr>
<td>Physio CRP</td>
<td>1.06 (1.01, 1.11)</td>
</tr>
<tr>
<td>Onset Sx (yrs)</td>
<td>1.02 (0.93, 1.12)</td>
</tr>
<tr>
<td>CTfib score ≥2</td>
<td>0.77 (0.29, 2.04)</td>
</tr>
</tbody>
</table>

Communication among multidisciplinary team members is essential for an accurate diagnosis.
Clinical Tools for Diagnosis

- Raise suspicion that ILD is present
- Identify the cause of the disease
  - Infection
  - Systemic disorders
  - Exposures (eg, occupational, environment, hobby)
  - Idiopathic

Clinical
- History and physical
- PFT
- Lab
Radiographic Tools for Diagnosis

HRCT: allows detailed evaluation of the lung parenchyma

HRCT Features
- Ground glass attenuation
- Honeycombing/cysts
- Lines/reticular thickening
- Consolidation
- Nodules
- Decreased lung attenuation

HRCT Distribution
- Upper
- Lower
- Central
- Peripheral
- Diffuse/bilateral
Histologic Tools for Diagnosis

**Histology**
- Bronchoscopy
- Surgical lung biopsy

**UIP Pattern**
- Marked fibrosis/architectural distortion ± honeycombing, predominantly subpleural/paraseptal
- Patchy fibrosis
- Fibroblastic foci
- Absence of features to suggest alternative diagnosis

1. Images courtesy of Steven Nathan, MD.
Usual Interstitial Pneumonia (UIP)

Idiopathic Pulmonary Fibrosis (IPF)

Rheumatoid Lung

Chronic Exposures
- Hypersensitivity pneumonia
- Occupational

Nonspecific Interstitial Pneumonia (NSIP)

Idiopathic

- Connective Tissue Disease
- Hypersensitivity Pneumonia

Organizing Pneumonia

Idiopathic COP/BOOP

OP due to:
- a very long list....
Causes of OP

Table 1. Causes of SOP

Associated with connective tissue disorders
- Systemic lupus erythematosus
- Rheumatoid arthritis
- Sjogren syndrome
- Polymyositis-dermatomyositis
- Polymyalgia rheumatica
- Systemic sclerosis
- Behcet’s disease
- Ankylosing spondylitis
- Mixed connective tissue disease

Associated with immunological disorders
- Common variable immunodeficiency syndrome
- Essential mixed cryoglobulinemia

Associated with infectious disease
Bacterial
- Streptococcus pneumoniae
- Legionella pneumophila
- Mycoplasma pneumoniae
- Coxiella burnetti
- Nocardia asteroides
- Chlamydia pneumoniae
- Staphylococcus aureus
Viral
- Adenovirus
- Cytomegalovirus
- Influenza and parainfluenza
- Human immunodeficiency virus
- Herpes virus
Fungal
- Cryptococcus neoformans
- Pneumocystis jiroveci
Parasites
- Plasmodium vivax

Associated with aspiration pneumonia

Associated with radiation therapy for breast cancer

Associated with organ transplantation
- Bone marrow
- Lung
- Renal
- Liver

Drug-related (see Table 2)

Miscellaneous
- Inflammatory bowel disease
- Primary biliary cirrhosis
- Polyarteritis nodosa
- Chronic thyroiditis
- Hematological malignancies (myelodysplastic syndrome, T-cell leukemia, lymphoma)
- Coronary artery bypass graft surgery
- Environmental exposure (textile printing dye, house fire, cocaine abuse)
- Sweet’s syndrome

Table 2. Drug-Associated OP

Most common:
- Amiodarone, bleomycin, carbamazepine, interferon-a, -b, gold salts

Less common:
- Acebutolol, doxorubicin, mesalamine, sulphasalazine, nitrofurantoin, sirolimus

Rare:
- Amphotericin B, bucillamine, busulfan, chlorambucil, cefradin, erlotinib, fluvastatin, L-tryptophan, minocycline, nilutamide, phenytoin, risedronate, rituximab, tacrolimus, temozolomide, thalidomide, ticlopidine, trastuzumab, vinbarbital

Adapted from Pneumotox (www.pneumotox.com).

High Resolution Computed Tomography

- Does NOT use contrast
- Thin collimation
  - HRCT, approximately 1mm slice thickness
  - MDCT (contiguous slices) preferred
    - Close tracking of subtle parenchymal and airway abnormalities
    - Avoids missing small/subtle abnormalities
- Should use Low Dose (~80 mA)
- Reconstruction with specific Windows
- Inspiration, Expiration, and prone images
High Resolution Computed Tomography

- Examines the entire lungs
  - Avoids sampling error (like surgical biopsy)
  - Can visualize mixed disease patterns
- Expiratory images add physiologic element
- Key Limitation is resolution
  - Ground Glass may be inflammation, fibrosis, infection, water, blood, etc.
  - Microscopic honeycomb change
  - Histopathologic features
Impact of Thickness & Algorithm

CT
10-mm standard algorithm

HRCT
1.5-mm high resolution algorithm
HRCT Pitfalls

- Dependent atelectasis mimics ground glass opacity
  - More common in smokers and with increased age
  - Always do prone images
Dependent Opacity: Normal

supine

prone
Dependent Opacity: Disease

supine

prone
Normal HRCT

- Clear 1 cm periphery
- Few interlobular septa
- Should see no airways in the peripheral 1/3 of the lungs; bronchioles not visible
- Dependent opacity
Mosaic Attenuation
(aka mosaic perfusion)

- wedge-shaped areas of alternating attenuation
- altered perfusion
  - pulmonary emboli
- altered ventilation
  - air-trapping
  - small airway disease
- patchy ground glass (ILD)
Inspiratory/Expiratory HRCT

Inspiration

Expiration
Emphysema vs. Cyst
Ground Glass

- Hazy opacity you can see through
  - Less opaque than consolidation
  - Able to see bronchial & vascular markings

- Partial filling of airspaces
  - Fluid (water, blood)
  - Infection
  - Fibrosis
Honeycombing

- Clustered cystic air spaces
- Well–defined walls
- Usually comparable diameter (3-10mm)
- Usually subpleural
- Can be confused with traction bronchiectasis
Respiratory Bronchiolitis / ILD

Pattern:
- Ill defined centrilobular nodules
- Ground Glass
- Decreased lobular attenuation

Distribution:
- mid/upper lungs
Langerhans Histiocytosis (aka EG)

Pattern:
- Numerous cysts (often bizarre shapes)
- Peribronchiolar nodules
- Interstitial changes/scar

Distribution:
- Upper lobe

Progression:
- Nodules $\rightarrow$ cavitary nodules $\rightarrow$ cysts $\rightarrow$ confluent cysts
Sarcoidosis

Pattern:
- Nodules
- Confluent alveolar spaces
- Distortion, fibrosis, cysts

Distribution:
- Upper lobe
- Central/bronchovascular
Lymphangioleiomyomatosis

Pattern:
- Numerous thin-walled cysts
- No nodules or fibrosis

Distribution:
- Diffuse, no predominance
Lymphangioleiomyomatosis
Hypersensitivity Pneumonitis

Pattern:
- Ground Glass
- Mosaic attenuation
- Peribronchiolar thickening

Distribution:
- Upper / Diffuse
50 year old male with Hypersensitivity Pneumonia – Treated with removal of doves and immunosuppression

<table>
<thead>
<tr>
<th></th>
<th>06/18/13</th>
<th>06/20/16</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1 (% pred)</td>
<td>1.95 (50%)</td>
<td>2.93 (78%)</td>
</tr>
<tr>
<td>FVC (% pred)</td>
<td>2.04 (38%)</td>
<td>3.22 (61%)</td>
</tr>
<tr>
<td>DLCO (% pred)</td>
<td>12.25 (38%)</td>
<td>25.22 (81%)</td>
</tr>
</tbody>
</table>

04/05/13 | 06/20/16
Hypersensitivity Pneumonitis - Chronic
Nonspecific interstitial pneumonia

Pattern:
• Reticulation
• Traction Bronchiectasis
• Ground Glass
• Honeycomb rare (5%)

Distribution:
• Lower
• Peripheral / Diffuse
Updated Consensus Statement for Diagnosis of IPF

The diagnosis of IPF requires:

1. Exclusion of other known causes of interstitial lung disease
2. Presence of UIP pattern on HRCT (in patients without surgical biopsy)
3. A HRCT pattern of definite/possible UIP with a Surgical lung biopsy showing Definite/Probable UIP

The Major and Minor Criteria proposed in the 2000 ATS/ERS Consensus Statement were Eliminated

Raghu et al., *Am J Respir Crit Care Med* 2011; 183:788-24
# Role of HRCT in Diagnosing UIP

<table>
<thead>
<tr>
<th>UIP Pattern (All 4 Features)</th>
<th>Possible UIP (All 3 Features)</th>
<th>Inconsistent With UIP (Any)</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Subpleural, basal predominance</td>
<td>• Upper or mid-lung predominance</td>
<td>• Upper or mid-lung predominance</td>
</tr>
<tr>
<td>• Reticular abnormality</td>
<td>• Peribronchovascular predominance</td>
<td>• Peribronchovascular predominance</td>
</tr>
<tr>
<td>• Honeycombing with/without traction bronchiectasis</td>
<td>• Extensive ground glass abnormality (extent &gt; reticular abnormality)</td>
<td>• Extensive ground glass abnormality (extent &gt; reticular abnormality)</td>
</tr>
<tr>
<td>• Absence of features listed as inconsistent with UIP (column 3)</td>
<td>• Profuse micronodules (bilateral, predominantly upper lobe)</td>
<td>• Profuse micronodules (bilateral, predominantly upper lobe)</td>
</tr>
<tr>
<td></td>
<td>• Discrete cysts (multiple, bilateral, away from areas of honeycombing)</td>
<td>• Discrete cysts (multiple, bilateral, away from areas of honeycombing)</td>
</tr>
<tr>
<td></td>
<td>• Diffuse mosaic attenuation/air-trapping (bilateral, in ≥3 lobes)</td>
<td>• Diffuse mosaic attenuation/air-trapping (bilateral, in ≥3 lobes)</td>
</tr>
<tr>
<td></td>
<td>• Consolidation in bronchopulmonary segment(s)/lobe(s)</td>
<td>• Consolidation in bronchopulmonary segment(s)/lobe(s)</td>
</tr>
</tbody>
</table>

Usual Interstitial Pneumonia

Pattern:
- irregular septal lines & honeycombing
- ground glass opacity (not predominant)
- traction bronchiectasis

Distribution:
- lower > upper lung
- subpleural distribution
## Radiology (HRCT) Diagnosis of IPF/UIP Versus NSIP

### Methodology
Consecutive patients with UIP or NSIP
- **n = 96**

### Results

<table>
<thead>
<tr>
<th>Diagnosis Type</th>
<th>HRCT Definite/Probable UIP</th>
<th>HRCT Not UIP</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>UIP</strong></td>
<td><strong>n = 27 (28%)</strong></td>
<td><strong>n = 69 (72%)</strong></td>
</tr>
<tr>
<td><strong>Non-UIP</strong></td>
<td><strong>n = 0 (0%)</strong></td>
<td></td>
</tr>
</tbody>
</table>

- **63% of UIP cases**

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Emphysema + IPF/UIP

<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>FVC</td>
<td>3.63 (89%)</td>
</tr>
<tr>
<td>FEV₁</td>
<td>2.74 (102%)</td>
</tr>
<tr>
<td>FEV₁/FVC</td>
<td>115%</td>
</tr>
<tr>
<td>RV</td>
<td>2.67 (113%)</td>
</tr>
<tr>
<td>TLC</td>
<td>6.30 (98%)</td>
</tr>
<tr>
<td>DLCO</td>
<td>11.90 (48%)</td>
</tr>
</tbody>
</table>
UIP: Irregular Reticular Opacities

Courtesy of W. Richard Webb, MD.
Early HRCT Findings in IPF

Courtesy of David A. Lynch, MD.
Histologic Tools for Diagnosis

**Histology**
- Bronchoscopy
- Surgical lung biopsy

**UIP Pattern**
- Marked fibrosis/architectural distortion ± honeycombing, predominantly subpleural/paraseptal
- Patchy fibrosis
- Fibroblastic foci
- Absence of features to suggest alternative diagnosis

1. Images courtesy of Steven Nathan, MD.
Idiopathic Pulmonary Fibrosis

A specific form of chronic, progressive fibrosing interstitial pneumonia of unknown cause, occurring primarily in older adults, and limited to the lungs.

It is characterized by progressive worsening of dyspnea and lung function and is associated with a poor prognosis.

Raghu et al., *Am J Respir Crit Care Med* 2011; 183:788-24
Having a Conversation With the Patient Newly Diagnosed With IPF

- Spend adequate time to explain the prognosis and assess patient's preferences and values
- Burden and morbidity of IPF can be emotionally overwhelming and will likely impact family members as well
- Each individual patient with IPF is different; consider physiology, exercise tolerance, radiology, and pathology when choosing a course of treatment
- Patients who are at increased risk of mortality should be referred for lung transplantation early in the course
## 2015 Treatment Recommendations for IPF

<table>
<thead>
<tr>
<th><strong>Strong Recommendation Against Use:</strong></th>
<th></th>
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<tr>
<td>Anticoagulation (warfarin), Pred/Aza/NAC, ambrisentan, Imatinib</td>
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<td>Nintedanib, pirfenidone, GERD</td>
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</tr>
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<tbody>
<tr>
<td>NAC, macitentan, bosentan, sildenafil</td>
<td></td>
</tr>
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</table>

High Dose Acetylcysteine in Idiopathic Pulmonary Fibrosis

Mortality
NAC = 9%
Placebo = 11%
p=0.69

Figure 2. Vital Capacity and Single-Breath Carbon Monoxide Diffusing Capacity (Dlco) at 6 and 12 Months, as Compared with Baseline.

Demedts et al; NEJM 2005;353:2229-42
PANTHER
Prednisone-Azathioprine-N-acetyl cysteine: A Trial That Evaluates Responses in IPF

Diagnosis of IPF with FVC ≥ 50%, DLCO ≥ 30% predicted

Three arms
- Placebo
- N-acetyl cysteine
- Pred/aza/NAC

Primary Endpoint – Change in FVC over 60wks
Interim Analysis with 50% data
- Combination n = 77, Placebo n= 78
- Increased Death 8 vs 1, p=0.01
- Increased Hosp 23 v 7, p<0.001
- No physio/clinical benefit

Termination of combination therapy at mean of 32 weeks
Recommendation against use of pred/azathioprine/N-acetyl cysteine
NAC Does Not Reduce FVC Decline

2015 Treatment Recommendations for IPF

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Nintedanib: INPULSIS-1 and INPULSIS-2 Trial Design

Inclusion Criteria
- Age ≥40
- IPF ≤5 y
- ≥50% FVC pred
- 30%-79% DLCO pred
- HRCT within 1 y

Endpoints
- $1^0$: Δ FVC
- $2^0$: Time to first AE
  Δ SGRQ

1,066 Patients

52 Weeks

3

Nintedanib
300 mg daily

2

Placebo

INPULSIS Primary Endpoint: Adjusted Annual Rate of Decline in FVC

**INPULSIS-1**
- **Nintedanib, 150 mg Twice Daily (N = 309)**: -114.7 ml/y
- **Placebo (N = 204)**: -239.9 ml/y

Difference, 125.3 (95% CI, 77.7-172.8)  
*P* < .001

**INPULSIS-2**
- **Nintedanib, 150 mg Twice Daily (N = 329)**: -113.6 ml/y
- **Placebo (N = 219)**: -207.3 ml/y

Difference, 93.7 (95% CI, 44.8-142.7)  
*P* < .001

INPULSIS: Time to First Investigator-Reported Acute Exacerbation

INPULSIS-1

Cumulative Incidence of First Investigator-Reported Acute Exacerbation, %

Day

HR 1.15 (95% CI, 0.54-2.42)
P = .67

Nintedanib, 150 mg Twice Daily
Placebo

INPULSIS-2

Cumulative Incidence of First Investigator-Reported Acute Exacerbation, %

Day

HR 0.38 (95% CI, 0.19-0.77)
P = .005

Placebo
Nintedanib, 150 mg Twice Daily

Nintedanib – Time to First Exacerbation Statified by FVC +/- 70% predicted
## Nintedanib – Safety & Tolerability

<table>
<thead>
<tr>
<th></th>
<th>Nintedanib (n=638)</th>
<th>Placebo (n=423)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dose Reduction*</td>
<td>178 (28%)</td>
<td>16 (4%)</td>
</tr>
<tr>
<td>Treatment Interruptions*</td>
<td>151 (24%)</td>
<td>42 (10%)</td>
</tr>
<tr>
<td></td>
<td>Incidence/Discontinue</td>
<td>Incidence/Discontinue</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>63% / 4.4%</td>
<td>18% / 0.2%</td>
</tr>
<tr>
<td>Nausea</td>
<td>25% / 2.0%</td>
<td>7% / 0%</td>
</tr>
<tr>
<td></td>
<td>Mild/Mod/Severe (%)</td>
<td>Mild/Mod/Severe (%)</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>57 / 38 / 5</td>
<td>77 / 20 / 3</td>
</tr>
<tr>
<td>Nausea</td>
<td>74 / 24 / 2</td>
<td>93 / 7 / 0</td>
</tr>
</tbody>
</table>

* No particular time
FDA Approval of Nintedanib

Approved October 15, 2014, for the treatment of IPF

Liver function tests required prior to treatment and should be evaluated every 3 months in first year

Dosage and administration

150 mg twice daily with food
Take each dose approximately 12 h apart

Adverse reactions? Consider temporary dose reduction to 100 mg, temporary interruption, or discontinuation
Pirfenidone: ASCEND Trial Design

**Inclusion Criteria**
- Age 40-80 y
- Confirmed IPF
- 50%-90% FVC pred
- 30%-90% DLCO pred
- FEV₁/FVC ≥0.80
- 6MWD ≥150 m

**Endpoints**
1. \( \Delta \) FVC or death
2. 6MWD
   - PFS
   - Dyspnea
   - Death

**52 Weeks**
- Pirfenidone 2,403 mg daily
- Placebo

**555 Patients**

ASCEND: Primary Efficacy Analysis

**Absolute difference**
- Week 13: 2.5%
- Week 26: 7.9%
- Week 39: 12.3%
- Week 52: 15.3%

**Relative difference**
- Week 13: 54.0%
- Week 26: 58.0%
- Week 39: 57.8%
- Week 52: 47.9%

**Rank ANCOVA P**
- Week 13: < .001
- Week 26: < .001
- Week 39: < .001
- Week 52: < .001

**ANCOVA**: analysis of covariance.

## Pirfenidone: Meta Analysis

### Table 2. Summary of finding form Pirfenidone for idiopathic pulmonary fibrosis.

<table>
<thead>
<tr>
<th>Outcomes</th>
<th>Anticipate absolute effects (Study population) (95% CI)</th>
<th>Relative Effect</th>
<th>NO of participants</th>
<th>Quality of the evidence (GRADE)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Risk with placebo</td>
<td>Risk with Pirfenidone</td>
<td>RR 0.53 (0.32 to 0.88)</td>
<td>1247 (3 RCTs)</td>
</tr>
<tr>
<td>All cause-mortality</td>
<td>67 per 1000</td>
<td>36 per 1000 (22 to 59)</td>
<td>RR 0.53 (0.32 to 0.88)</td>
<td>1247 (3 RCTs)</td>
</tr>
<tr>
<td></td>
<td>442 per 1000</td>
<td>372 per 1000 (332 to 416)</td>
<td>RR 0.83 (0.75 to 0.94)</td>
<td>728 (3 RCTs)</td>
</tr>
<tr>
<td>Progression free-survival</td>
<td>26 per 1000</td>
<td>15 per 1000 (5 to 47)</td>
<td>RR 0.59 (0.19 to 1.84)</td>
<td>235 (2 RCTs)</td>
</tr>
<tr>
<td>Acute exacerbation</td>
<td>168 per 1000</td>
<td>107 per 1000 (84 to 139)</td>
<td>RR 0.64 (0.50 to 0.83)</td>
<td>1615 (5 RCTs)</td>
</tr>
<tr>
<td>Worsening of IPF</td>
<td>417 per 1000</td>
<td>308 per 1000 (267 to 358)</td>
<td>RR 0.74 (0.64 to 0.86)</td>
<td>1236 (3 RCTs)</td>
</tr>
<tr>
<td>Change on 6MWT</td>
<td>30 per 1000</td>
<td>68 per 1000 (40 to 115)</td>
<td>RR 2.26 (1.33 to 3.83)</td>
<td>764 (5 RCTs)</td>
</tr>
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</table>

1: Non primary outcome from RCTs, 2: High heterogeneity; 6MWT: Six minutes walk test; RCT: Randomized controlled trial; RR: Risk ratio; CI: confidence interval.

ASCEND: Treatment-Emergent Adverse Events more common in pirfenidone group

- Nausea (36% vs 13%)
- Rash (28% vs 9%)

- Adverse events (AEs) generally mild to moderate severity, reversible, and without clinically significant sequelae

FDA Approval of Pirfenidone

Approved October 15, 2014, for the treatment of IPF

Liver function tests required prior to treatment and should be evaluated every 3 months in first year

Dosage and administration

- 801 mg 3x daily with food (three 267-mg capsules per dose)
- Take each dose at the same time each day
- Initiate with titration
  - Days 1-7: one capsule 3x daily
  - Days 8-14: two capsules 3x daily
  - Days 15 onward: three capsules 3x daily

Adverse reactions? Consider temporary dosage reduction, treatment interruption, or discontinuation
Gastroesophageal reflux (GERD) in IPF

- GER is highly prevalent in patients with IPF
- Observational study (n = 204); 47% received GER medical therapy, and 5% surgical
Engaging in a Shared Decision-Making Process

- Discuss the efficacy and safety of FDA-approved therapies
- Listen to patient’s preferences and concerns
- Focus on symptom control and management of comorbidities
- Set treatment expectations
- Look at the option of lung transplantation

Physician provides
- Treatment options
- Risks and benefits

Mutually acceptable decision

Patient provides
- Personal preferences
- Values and concerns
Members of the IPF Care Team

- Multidisciplinary Team of Physicians
  - Pulmonary, Radiology, Pathology, Rheumatology, Cardiology, Thoracic Surgery, Lung Transplant
- Social Work
- Clinical Nurse Specialist
- Palliative Care
- Students/Residents/Fellows
- Research Coordinator
- Support Group
Supportive Care for Patients With IPF

Supportive Care Options

- Educate patients
  - Refer to reliable sources

- Prescribe O₂
  - Screen for resting/nocturnal/exertional requirement

- Close monitoring of symptoms and pulmonary function

- Exercise
  - Pulmonary rehabilitation

- Treatment of comorbid illness
  - GERD, OSA, CAD

OSA: obstructive sleep apnea.
Lung Transplantation for Pulmonary Fibrosis: Referral and Listing Guidelines

Referral

- Diagnosis of IPF (histologic or radiographic)
- Diagnosis of fibrotic NSIP (histologic)

Transplantation

- $\text{DL}_\text{CO} < 39\%$ predicted
- Decline in FVC by $\geq 10\%$ over 6 months
- Oxyhemoglobin saturation $< 88\%$ with 6MWT
- Honeycombing on HRCT
- Histologic evidence of NSIP and
  - $\text{DL}_\text{CO} < 35\%$ predicted
  - Decline in FVC of $\geq 10\%$ over 6 months
  - Decline in $\text{DL}_\text{CO}$ of $\geq 15\%$ over 6 months

www.pulmonaryfibrosis.org

Serving the PF Community
Call 844.TalkPFF (844.825.5733) to speak with a representative today. LEARN MOI

We Imagine a World Without Pulmonary Fibrosis