Idiopathic Pulmonary Fibrosis - What You Need to Know: A Case-based update on diagnosis and treatment

Emerging Challenges in Primary Care: Update 2013

Faculty

• Kevin R. Flaherty, MD, MS
  - Associate Professor, Pulmonary and Critical Care Medicine, University of Michigan Health System, Ann Arbor, MI

• Fernando Martinez, MD, MS
  - Professor Department of Internal Medicine, Director Pulmonary Diagnostic Service, Associate Division Chief for Clinical Research, University of Michigan, Ann Arbor, MI
Faculty Disclosure

- Kevin R. Flaherty, MD, MS
  - Advisory Board – Boehringer Ingelheim, Medimmune, Gilead, Fibrogen, GlaxoSmithKline, Genentech, Ikaria, Novartis, Takeda, Vertex
  - Speaking - Boehringer Ingelheim, Pfizer, Forest, GlaxoSmithKlein

- Fernando Martinez, MD, MS
  - Advisory Board – Biogen, Forest, GlaxoSmithKline, Ikaria, Janssens, Merck, Nycomed/Takeda, Stromedix, Vertex
  - Royalties - Informa
  - Steering Committee – Forest, GlaxoSmithKline, Nycomed/Takeda
  - Speaker’s Bureau – Bayer, Forest, GlaxoSmithKline, Nycomed/Takeda
  - Teleconferences/CME Programs – American Institute for Research, CME Incite, Grey, MedScape, NACE, NACME, Projects in Knowledge, Sudler & Hennessey, UIC, UTSW, UptoDate, WSU

Learning Objectives

- Explain the basic pathobiology of IPF to patients
- Apply a state-of-the-art approach to diagnosing idiopathic pulmonary fibrosis (IPF)
- Define prognostic features for individual IPF patients
- Recognize the role of available non-pharmacological therapies including pulmonary rehabilitation, oxygen supplementation and lung transplantation in IPF management
- Apply appropriate pharmacotherapeutic options for individual IPF patients
- Develop a general understanding of novel treatments under intense investigation
On a scale of 1 to 5, please rate how confident you would be in discussing the diagnostic and treatment strategies for a patient with Idiopathic Pulmonary Fibrosis.

1. Not at all confident
2. Slightly confident
3. Moderately confident
4. Pretty much confident
5. Very confident

A 70 yo male presents with 12 months of progressive cough and breathlessness. After initial history, physical, laboratory studies and pulmonary function testing what high resolution chest computed tomography features will ensure a diagnosis of usual interstitial pneumonia (UIP)?

1) Basilar predominance of abnormality
2) Honeycomb change
3) Neither
4) 1 & 2
In this 70 yo male with a HRCT picture of UIP, which of the following features would exclude a diagnosis of idiopathic pulmonary fibrosis (IPF)?

1) Collagen vascular illness
2) Absence of occupational exposure
3) Cardiovascular co-morbidity
4) DL\(_{CO}\) > 50% predicted

In this 70 yo male which of the following features obtained at the time of diagnosis would suggest(s) a worse prognosis?

1) Age > 65 years
2) FVC > 50% predicted
3) DL\(_{CO}\) > 35% predicted
4) All of the above
In this 70 yo male with confirmed diagnosis of IPF and exertional desaturation, which of the following therapies should be considered?

1) Azathioprine, prednisone, N-acetyl cysteine
2) Prednisone alone
3) Inhaled corticosteroids
4) None of the above

58 yo male with mild cough/dyspnea

HPI
Several months of mild exertional breathlessness
Increased non-productive cough
No systemic symptoms
Prior CXR with ‘mild scarring’
Seen in Urgent Care three weeks ago and treated for ‘walking pneumonia’ based on CXR

EXAM
BP 148/84, P 90, RR 18, T 98.6
Basilar rales
No JVD
Normal CV
Normal abdomen
No joint/skin findings
Nonfocal neuro exam

PMH
Hypercholesterolemia - simvastatin
BPH – doxazosin
SH – Never Smoker, Engineer w/o exposures
What would be your first diagnostic impression?

1. Atypical pneumonia
2. Congestive heart failure
3. COPD
4. Interstitial lung disease
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Interstitial Lung Diseases - Difficulties

- Diverse group of disorders (130+)
- Similar symptoms, physiology, radiology
- Difficult nomenclature
- Limited, often toxic, treatments
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.

Five year survival of IPF is worse than most cancers

![Graph showing five year survival rate for IPF and different cancers](image-url)
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

Raghu et al., *Am J Respir Crit Care Med* 2011; 183:788-24

Classification of Diffuse Parenchymal Pulmonary Disorders

Diffuse parenchymal lung disease (DPLD)

DPLD of known cause

Idiopathic Interstitial pneumonia

Granulomatous DPLD

Other DPLD

Idiopathic UIP = IPF

Non-UIP IIP

DIP

RBILD

AIP

COP (BOOP)

NSIP

LIP

Classification of Diffuse Parenchymal Pulmonary Disorders

Diffuse parenchymal lung disease (DPLD)

DPLD of known cause

Idiopathic Interstitial pneumonia

Granulomatous DPLD

Other DPLD

Idiopathic UIP = IPF

Non-UIP IIP

DIP

RBILD

AIP

COP (BOOP)

NSIP

LIP

Question 1: Is DPLD possible?

Question 2: Is it idiopathic?

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**Classification of Diffuse Parenchymal Pulmonary Disorders**

- Diffuse parenchymal lung disease (DPLD)
  - DPLD of known cause
  - Idiopathic Interstitial pneumonia
  - Granulomatous DPLD
  - Other DPLD

**Question 3: Is it idiopathic UIP?**

- Idiopathic UIP = IPF

**Flowchart: Patient with suspected ILD**

- Hx, PE, CXR, PFT, Labs
- Dx likely by bronch?
- Yes: Is bronch diagnostic? Yes
- No: HRCT
  - Hx & HRCT consistent with IPF
    - STOP
  - Hx & HRCT Dx of other ILD
    - STOP
  - Suspected other ILD
    - Atypical clinical or CT features of IPF
      - Dx likely by bronch?
        - Yes
          - Is bronch diagnostic? Yes
          - Surgical bx
        - No
          - STOP
        - STOP
      - No
        - Surgical bx

**References**

- Noth & Martinez, Chest 2007; 132: 637-50
Idiopathic Pulmonary Fibrosis - What You Need to Know: A Case-based update on diagnosis and treatment

**Diagnostic “Tools”**

**Clinical**
- History & Physical, PFT, Lab

1. Raise suspicion that ILD is present
2. Identify a cause of the disease
   a. Infection
   b. Systemic Disorders
   c. Exposures (inhaled or oral)
   d. Idiopathic

**UIP Associated With Collagen Vascular Disease (CVD) Is Associated With Improved Prognosis and is not considered idiopathic pulmonary fibrosis**

Flaherty et al; AJRCCM 2003; 167: 1410-5

Park et al; AJRCCM 2007; 175: 705-11

NACE - Emerging Challenges in Primary Care: 2013
ILD: Laboratory screen for collagen vascular disease

- Creatinine phosphokinase
- Anti-nuclear antibody
- Extractable nuclear antibody panel

Pulmonary Function Testing

- Pulmonary Mechanics – FEV₁, FVC, FEV₁/FVC
  - Obstructive Lung Disease → Decreased FEV₁/FVC ratio
  - Restrictive Lung Disease → Normal/Increased FEV₁/FVC ratio
  - Muscle weakness → Normal/Increased FEV₁/FVC ratio
  - Percent predicted grades severity of FEV₁ and FVC
- Lung Volumes
  - True measure of size of lung
  - Total lung capacity (TLC), residual volume (RV)
- Diffusion capacity for carbon monoxide (DL_CO)
  - Decreased in many diseases such as emphysema, interstitial lung diseases, pulmonary vascular disease, pulmonary emboli
### Restrictive lung disease

- Which of the following does not cause an obstructive pattern?
  1. COPD
  2. Asthma
  3. Interstitial lung disease
  4. None of the above
60 year old smoker with progressive SOB

• What is your leading general diagnosis?
  1. COPD
  2. Interstitial lung disease
  3. Obesity
  4. Congestive heart failure
**Idiopathic Pulmonary Fibrosis - What You Need to Know: A Case-based update on diagnosis and treatment**

### Patient with suspected ILD

- **Hx, PE, CXR, PFT, Labs**
  - **Dx likely by bronch?**
    - Yes
    - **Is bronch diagnostic?**
      - Yes
        - **HRCT**
          - **Hx & HRCT consistent with IPF**
            - STOP
          - **Hx & HRCT Dx of other ILD**
            - STOP
        - No
          - **Suspected other ILD**
            - **Atypical clinical or CT features of IPF**
              - **Dx likely by bronch?**
                - Yes
                  - **Is bronch diagnostic?**
                    - Yes
                      - **Surgical bx**
                    - No
                      - STOP
                - No
                  - STOP

### Diagnostic “Tools”

**Radiographic**

- **CXR, HRCT**

**HRCT Features**

- Ground glass attenuation
- Honeycombing/cysts
- Lines/Reticular thickening
- Consolidation
- Nodules
- Decreased lung attenuation

**HRCT Distribution**

- Upper
- Lower
- Central
- Peripheral
- Diffuse/Bilateral
High Resolution Computed Tomography

Allows detailed evaluation of the *lung parenchyma*

Optimal for *interstitial lung disease*, infection, emphysema, bronchiectasis

Technique

- Does NOT use contrast
- Thin collimation with approximately 1mm slice thickness
- Reconstruction with specific Windows
- Inspiration, Expiration, and prone images

- *Regular CT or PE CT for everything else*

**UIP features**

- Upper lobes
- Irregular Lines
- Peripheral/Subpleural
- Honeycomb
- Lower lobes
- Lower lobe predominant

34
Usual Interstitial Pneumonia is lower lobe predominant

Accuracy of diagnosis of UIP

<table>
<thead>
<tr>
<th>Study</th>
<th>Correctness of first choice diagnosis</th>
<th>Correctness of confident first choice</th>
<th>% of UIP cases with confident diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mathieson</td>
<td>89%</td>
<td>95%</td>
<td>72%</td>
</tr>
<tr>
<td>Lee</td>
<td>88%</td>
<td>100%</td>
<td>71%</td>
</tr>
<tr>
<td>Swensen</td>
<td>89%</td>
<td>100%</td>
<td>67%</td>
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<tr>
<td>Hunninghake</td>
<td>85%</td>
<td>96%</td>
<td>48%</td>
</tr>
</tbody>
</table>
Diagnosis of Interstitial Lung Disease (ILD)

1. **Patient with suspected ILD**
2. **Hx, PE, CXR, PFT, Labs**
   - **Dx likely by bronch?**
     - Yes
     - Is bronch diagnostic?
       - Yes
       - STOP
     - No
       - HRCT
         - Hx & HRCT consistent with IPF
           - STOP
         - Hx & HRCT Dx of other ILD
           - STOP
         - Suspected other ILD
           - Atypical clinical or CT features of IPF
             - Dx likely by bronch?
               - Yes
               - Is bronch diagnostic?
                 - Yes
                 - STOP
               - No
                 - Surgical bx
                   - UIP
                   - NSIP
                   - RBILD
                   - DIP
                   - DAD
                   - OP
                   - LIP
                   - Non IIP

Diagnostic Thoracoscopic Biopsy: An Outpatient Experience

- 62 patients with ILD or pulmonary nodules
  - Chest tube removed in no air-leak and CXR without residual PTX

Results
- 45 (73%) discharged in 8 hours
- 14 (23%) discharged within 23 hours
  - Comorbidity (8), pain (4), air-leak (1), thoracotomy (1)
- 3 admitted
  - Prolonged air-leak (2), conversion to thoracotomy (1)

Chang et al, 2002 *Ann Thorac Surg* 74, 1942-6
The Clinical Radiographic and Pathologic Diagnosis of IIP:
Clinical Gold Standard

Multidisciplinary communication is essential to an accurate diagnosis

Raghu et al., *Am J Respir Crit Care Med* 2011; 183:788-24

**Learning Objectives**

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**Disease Progression in IPF**

is Variable and often Unpredictable

Disease Progression

Minimal Symptoms
Hypoxemia
Increased Disability
Pulmonary HTN
Death


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**A Multidimensional Index and Staging System for Idiopathic Pulmonary Fibrosis**

<table>
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<tr>
<th>Predictor</th>
<th>Points</th>
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<td>G (Gender)</td>
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<tr>
<td>Female</td>
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<td>Male</td>
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</tr>
<tr>
<td>Age (yr)</td>
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<td>&lt;60</td>
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<td>60-69</td>
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<tr>
<td>&gt;70</td>
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<tr>
<td>Physician</td>
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<tr>
<td>PVC % predicted</td>
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<td>&gt;80-75</td>
<td>1</td>
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<tr>
<td>&lt;80</td>
<td>2</td>
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<tr>
<td>Disease % predicted</td>
<td>0</td>
</tr>
<tr>
<td>&gt;95</td>
<td>1</td>
</tr>
<tr>
<td>70-90</td>
<td>2</td>
</tr>
<tr>
<td>&lt;70</td>
<td>3</td>
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<tr>
<td>Can't perform</td>
<td>3</td>
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Total Possible Points: 8

Stage | I | II | III |
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<tr>
<td>Points</td>
<td>0-3</td>
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<td>Mortality</td>
<td>5y</td>
<td>10y</td>
<td>15y</td>
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<td>9.6</td>
<td>9.0</td>
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<td>15.9</td>
<td>15.3</td>
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<td>39.3</td>
<td>29.9</td>
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<td>42.1</td>
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<td>76.8</td>
<td>76.8</td>
<td>76.8</td>
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</table>

Idiopathic Pulmonary Fibrosis - What You Need to Know: A Case-based update on diagnosis and treatment

Prognosis - Summary

• **Baseline factors** associated with ↑ risk for mortality
  – Older Age
  – Decreased FVC, DL_{CO}, 6 minute walk distance, VO_{2} max
  – Impaired oxygenation (rest and with exercise)
  – Desaturation during a 6-minute walk test (≤ 88%)
  – Presence of pulmonary hypertension
  – Failure of heart rate to recover after 6-minute walk test
  – Cough

• **Longitudinal factors** associated with ↑ risk for mortality
  – Acute Exacerbation/Respiratory Hospitalization
  – Decline in FVC of 5 - 10%
  – Decline in DL_{CO}
  – Increase in dyspnea
  – Decrease in walk distance/development of desaturation

• In many patients the course is still unpredictable

Exacerbations of IPF are not infrequent

461 patients with IPF (269 biopsy proven)

• 163 Respiratory Deteriorations requiring hospitalization
  – Focal x-ray lesion (pneumothorax, pneumonia) 14%,
  – Diffuse in 86%
  – Exacerbation in 55%
  – Infection 31% (opportunistic in 57%)
  – Heart Failure (3%)

<table>
<thead>
<tr>
<th></th>
<th>Acute Exacerbation</th>
<th>Respiratory Deterioration</th>
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</thead>
<tbody>
<tr>
<td>1 year</td>
<td>58 (14.2%)</td>
<td>97 (23%)</td>
</tr>
<tr>
<td>2 year</td>
<td>71 (18.8%)</td>
<td>124 (31.2%)</td>
</tr>
<tr>
<td>3 year</td>
<td>75 (20.7%)</td>
<td>134 (35.4%)</td>
</tr>
</tbody>
</table>

*Song et al., Eur Resp J 2011; 37:356-63*
Learning Objectives

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Patient with IPF

Best Supportive Treatment  Empiric Medical Treatment  Clinical Trial

Lung Transplant
IPF: ‘Supportive’ Treatment

- Close monitoring of symptoms and pulmonary function
- Treatment of Comorbid illness
  - ? GERD
  - ? Pulmonary Hypertension
- Exercise – Pulmonary Rehabilitation
- Oxygen

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An Official ATS/ERS/JRS/ALAT Statement: Idiopathic Pulmonary Fibrosis: Evidence-based Guidelines for Diagnosis and Management

Rationale:
Numerous studies that have enhanced our understanding of IPF have been published since the last IPF statement in 2000

Objective:
Provide an evidence based guideline on the diagnosis and management of IPF.
Provide recommendations using GRADE methodology

Raghu et al., Am J Respir Crit Care Med 2011; 183:788-24

**Evidence**

<table>
<thead>
<tr>
<th>Strength</th>
<th>Prednisone alone</th>
<th>NAC+AZA+Prednisone</th>
<th>Prednisone combined</th>
<th>NAC alone</th>
<th>Interferon-gamma</th>
<th>Anticoagulants</th>
<th>Bosentan</th>
<th>Pirfenidone</th>
<th>Cyclosporine A</th>
<th>Etanercept</th>
<th>Colchicine</th>
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</thead>
<tbody>
<tr>
<td>Weak</td>
<td>Prednisone alone</td>
<td>NAC+AZA+Prednisone</td>
<td>Prednisone combined</td>
<td>NAC alone</td>
<td>Interferon-gamma</td>
<td>Anticoagulants</td>
<td>Bosentan</td>
<td>Pirfenidone</td>
<td>Cyclosporine A</td>
<td>Etanercept</td>
<td>Colchicine</td>
</tr>
<tr>
<td>Strong</td>
<td>Prednisone alone</td>
<td>NAC+AZA+Prednisone</td>
<td>Prednisone combined</td>
<td>NAC alone</td>
<td>Interferon-gamma</td>
<td>Anticoagulants</td>
<td>Bosentan</td>
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<td>Cyclosporine A</td>
<td>Etanercept</td>
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</tbody>
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Raghu et al., Am J Respir Crit Care Med 2011; 183:788-24
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**Prednisone, Azathioprine, and N-Acetylcysteine for Pulmonary Fibrosis**

The Idiopathic Pulmonary Fibrosis Clinical Research Network

- 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


**Other Recent Negative Trials**

<table>
<thead>
<tr>
<th>Drug</th>
<th>Primary Endpoint</th>
<th>N</th>
<th>Trial length (weeks)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Imatinib mesylate</td>
<td>FVC drop 10% or death</td>
<td>119</td>
<td>96</td>
</tr>
<tr>
<td>Etanercept</td>
<td>FVC and DL$_{CO}$, % predicted, A-a gradient</td>
<td>88</td>
<td>48</td>
</tr>
<tr>
<td>Bosentan (BUILD 1)</td>
<td>Change 6MWT</td>
<td>158</td>
<td>52</td>
</tr>
<tr>
<td>Bosentan (BUILD 3)</td>
<td>Dz prog/Death/Exac</td>
<td>616</td>
<td>Events</td>
</tr>
<tr>
<td>Ambrisentan</td>
<td>Dz prog/Death/Resp Hosp</td>
<td>660</td>
<td>Events</td>
</tr>
<tr>
<td>Warfarin (ACE-IPF)</td>
<td>Death/drop FVC/non-elective hospitalization</td>
<td>248</td>
<td>48</td>
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<tr>
<td>Everolimus</td>
<td>2nd of FVC/TLC drop 10%, DLCO drop 15%, SaO$_2$-4%</td>
<td>104</td>
<td>156</td>
</tr>
</tbody>
</table>

Daniels et al., *Am J Resp Crit Care Med* 2010; 181:504-10
Raghu et al., 2008 *Am J Resp Crit Care Med* 178:948-55
King et al., 2008 *Am J Resp Crit Care Med* 177:75-81

Malouf et al., 2011 *Respirology* 2011;16:776-83
[www.clinicaltrials.gov](http://www.clinicaltrials.gov)
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**Change in FVC in CAPACITY (pirfenidone) Studies**

<table>
<thead>
<tr>
<th>Week</th>
<th>CAPACITY 1</th>
<th>CAPACITY 2</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>LS Mean Change</td>
<td>Rank ANCOVA</td>
</tr>
<tr>
<td></td>
<td>PFD</td>
<td>Placebo</td>
</tr>
<tr>
<td>72</td>
<td>-6.49</td>
<td>-7.23</td>
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*Change in FVC in CAPACITY (pirfenidone) Studies*


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**BIPF (Tyrosine Kinase Inhibitor)**

- 12 month study, 432 patients with IPF

*BIPF (Tyrosine Kinase Inhibitor)*

*Richeldi et al., New Eng J Med 2011;265:1079-87*
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Adamali H, Maher T Drug Des Devel Ther 2012:261-71

ONGOING CLINICAL TRIALS IN IPF*

<table>
<thead>
<tr>
<th>Product</th>
<th>Company</th>
<th>MOA</th>
<th>Phase</th>
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<tbody>
<tr>
<td>Pirfenidone</td>
<td>InterMune</td>
<td>Unknown</td>
<td>3</td>
</tr>
<tr>
<td>BIBF 1120</td>
<td>Boehringer Ingelheim</td>
<td>TKI</td>
<td>3</td>
</tr>
<tr>
<td>QAX 576</td>
<td>Novartis</td>
<td>Anti-IL13 MoAb</td>
<td>2</td>
</tr>
<tr>
<td>FG-3019</td>
<td>Fibrogen</td>
<td>Anti-CTGF MoAb</td>
<td>2</td>
</tr>
<tr>
<td>STX-100</td>
<td>Biogen/Stromedix</td>
<td>Anti-α,β6 integrin MoAb</td>
<td>2</td>
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<tr>
<td>CNTO 888</td>
<td>Centocor</td>
<td>Anti-CCL-2 MoAb</td>
<td>2</td>
</tr>
<tr>
<td>CC-930</td>
<td>Celgene</td>
<td>Jun-kinase inhibitor</td>
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<td>BMS-986202</td>
<td>Bristol-Myers Squibb</td>
<td>LPA1 receptor antagonist</td>
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<tr>
<td>ASY12295</td>
<td>Sanofi-Aventis</td>
<td>IL4/IL13 MoAb</td>
<td>2</td>
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<tr>
<td>IW001</td>
<td>ImmuneWorks</td>
<td>Type V collagen</td>
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<tr>
<td>PRM-151</td>
<td>Promedior</td>
<td>Human pentraxin</td>
<td>1</td>
</tr>
</tbody>
</table>

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*List likely incomplete
Timing for Referral for Lung Transplantation:  
Idiopathic Pulmonary Fibrosis

• Guideline for referral
  – Histologic or radiographic evidence of UIP irrespective of FVC
  – Histologic evidence of fibrotic NSIP

• Guideline for transplantation
  – DLCO < 39% predicted
  – 10% or greater decrement in FVC or 15% decrease in DLCO during 6 month follow up
  – Fall in SaO2 below 88% during a 6MW test
  – Honeycombing on HRCT

Orens et al. JHLT 2006;25:745

Delayed access to tertiary care among IPF patients is associated with increased mortality

IDiopathic Pulmonary Fibrosis - What You Need to Know: A Case-based update on diagnosis and treatment

**IPF - Summary**

- A form of chronic, progressive fibrosing interstitial pneumonia of unknown cause
- Occurs primarily in older adults and is limited to the lungs
- Diagnostic process is centered on:
  - Excluding systemic diseases or exposures
  - Identifying a pattern of UIP on HRCT or Surgical lung biopsy
- Standard immunosuppressive therapy is no longer felt to be indicated
- New and promising treatments are being studied

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**62 year old with breathlessness and cough**

**History**

- Several months of progressive SOB/cough
- Negative PMH
- SH
  - Non-smoker
  - Two parakeets at home

**Physical examination**

- HEENT – normal
- No adenopathy
- Basilar crackles
- CV – normal
- Abd - normal
62 year old with breathlessness and cough

What is your leading general diagnosis?
1. COPD
2. Hypersensitivity pneumonitis
3. Idiopathic pulmonary fibrosis (IPF)
4. Congestive heart failure
58 year old with several years of breathlessness

Mild cough

PMH
- Breast Cancer 3 years earlier
- Progressive systemic sclerosis (scleroderma)

Non-smoker

No exposures

60 year old smoker with progressive SOB

What is your leading general diagnosis?
1. COPD
2. Idiopathic pulmonary fibrosis (IPF)
3. Congestive heart failure
4. Connective tissue associated interstitial lung disease
A 70 yo male presents with 12 months of progressive cough and breathlessness. After initial history, physical, laboratory studies and pulmonary function testing what high resolution chest computed tomography features will ensure a diagnosis of usual interstitial pneumonia (UIP)?

1) Basilar predominance of abnormality
2) Honeycomb change
3) Neither
4) 1 & 2
In this 70 yo male with a HRCT picture of UIP, which of the following features would exclude a diagnosis of idiopathic pulmonary fibrosis (IPF)?

1) Collagen vascular illness
2) Absence of occupational exposure
3) Cardiovascular co-morbidity
4) DL$_{CO}$ > 50% predicted

In this 70 yo male which of the following features obtained at the time of diagnosis would suggest(s) a worse prognosis?

1) Age > 65 years
2) FVC > 50% predicted
3) DL$_{CO}$ > 35% predicted
4) All of the above
In this 70 yo male with confirmed diagnosis of IPF and exertional desaturation, which of the following therapies should be considered?

1) Azathioprine, prednisone, N-acetyl cysteine
2) Prednisone alone
3) Inhaled corticosteroids
4) None of the above

On a scale of 1 to 5, please rate how confident you would be in discussing the diagnostic and treatment strategies for a patient with Idiopathic Pulmonary Fibrosis.

1. Not at all confident
2. Slightly confident
3. Moderately confident
4. Pretty much confident
5. Very confident
How likely are you to implement strategies learned from this presentation in your practice?

1. Very likely
2. Somewhat likely
3. Unlikely
4. Not applicable