Evaluation and Treatment of Idiopathic Pulmonary Fibrosis

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Case

• 57 yo WM
• SOB over the past 6 months
• Throat clearing, dry cough for 3 years

• DOE at work, difficulty climbing steps
• Not feeling better after cath/PTCA 2 months prior
• Abnormal CXR showing fibrosis

• PMHx: CAD, GERD
• Meds: ASA, Plavix, metoprolol, PPI
• PSHx: 15PY tob, quit 20 years ago
Case

- Exam
  - Bibasilar dry crackles
  - Mild clubbing
- PFTs:
  - Normal spirometry
  - Lung volumes restriction TLC 68% predicted
  - DLCO 55% predicted
  - 6 minute walk: 2100 feet, 97% at rest, 84% with walk on room air
- Labs:
  - ANA (+) 1:80
  - RF (+), ANCA (-), ENA (-)
- CXR shows interstitial lung disease

Interstitial Lung Diseases

- Groups of disorders characterized by varying degrees of inflammation and fibrosis
- Response to a known tissue injury or part of unknown process
- Dysregulated repair process
- Effect the interstitial space
  - Between the alveolar epithelial cell membrane-pulmonary capillary endothelial cell membrane
  - Site of initial injury, early effects on gas transfer
### Interstitial Lung Diseases

- Can also affect areas outside the alveoli, such as the bronchioles, larger airways and pulmonary vasculature
  - Diffuse parenchymal lung diseases

### Interstitial Lung Disease

- Over 150 etiologies
- Symptoms nonspecific
  - SOB/DOE and cough
- Diagnosis requires combination of:
  - Clinical presentation
  - Radiology (high resolution chest CT)
  - Pathology
- Prognosis and treatment dependent on diagnosis
### Interstitial Lung Disease

<table>
<thead>
<tr>
<th>Conditions</th>
</tr>
</thead>
<tbody>
<tr>
<td>Desquamative Interstitial Pneumonitis</td>
</tr>
<tr>
<td>Lymphocytic Interstitial Pneumonitis</td>
</tr>
<tr>
<td>Eosinophilic pneumonia</td>
</tr>
<tr>
<td>Alveolar Proteinosis</td>
</tr>
<tr>
<td>Amyloidosis</td>
</tr>
<tr>
<td>Lymphangitic Carcinomatosis</td>
</tr>
<tr>
<td>Radiation Pneumonitis</td>
</tr>
<tr>
<td>Langerhan's Cell Granulomatosis</td>
</tr>
<tr>
<td>Lymphangioleiomyomatosis</td>
</tr>
<tr>
<td>Tuberous Sclerosis</td>
</tr>
<tr>
<td>Neurofibromatosis</td>
</tr>
<tr>
<td>Hypersensitivity Pneumonitis</td>
</tr>
<tr>
<td>Sarcoidosis</td>
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<tr>
<td>Berylliosis</td>
</tr>
<tr>
<td>Ankylosing Spondylitis</td>
</tr>
<tr>
<td>Rheumatoid Arthritis</td>
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<tr>
<td>Silicosis</td>
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<tr>
<td>Asbestosis</td>
</tr>
<tr>
<td>Lymphoma</td>
</tr>
<tr>
<td>Hemosiderosis</td>
</tr>
<tr>
<td>Wegener’s Granulomatosis</td>
</tr>
<tr>
<td>Drug-Induced Fibrosis</td>
</tr>
<tr>
<td>Systemic Sclerosis</td>
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<tr>
<td>Systemic Lupus Erythematosus</td>
</tr>
<tr>
<td>Sjogren’s Syndrome</td>
</tr>
<tr>
<td>Mycobacterial Infection</td>
</tr>
<tr>
<td>Histoplasmosis</td>
</tr>
<tr>
<td>Aspiration</td>
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<tr>
<td>Lipoid Pneumonia</td>
</tr>
<tr>
<td>Polymyositis</td>
</tr>
<tr>
<td>Mixed Connective Tissue Disease</td>
</tr>
<tr>
<td>Microlithiasis</td>
</tr>
<tr>
<td>Churg-Strauss Syndrome</td>
</tr>
<tr>
<td>Pneumocystis carinii</td>
</tr>
<tr>
<td>Oxygen Toxicity</td>
</tr>
<tr>
<td>Cryptogenic Organizing Pneumonia</td>
</tr>
<tr>
<td>Non-Specific Interstitial Pneumonitis</td>
</tr>
<tr>
<td>Usual Interstitial Pneumonitis</td>
</tr>
<tr>
<td>Bleomycin</td>
</tr>
<tr>
<td>IgG4 disease</td>
</tr>
<tr>
<td>Hard metal disease</td>
</tr>
<tr>
<td>Crohn’s disease</td>
</tr>
<tr>
<td>Ulcerative colitis</td>
</tr>
<tr>
<td>Idiopathic inflammatory myopathy</td>
</tr>
<tr>
<td>Familial IPF</td>
</tr>
<tr>
<td>Hermansky-Pudlak syndrome</td>
</tr>
<tr>
<td>Gaucher’s disease</td>
</tr>
<tr>
<td>Goodpasture’s syndrome</td>
</tr>
<tr>
<td>Nitrofurantoin</td>
</tr>
<tr>
<td>Methotrexate</td>
</tr>
<tr>
<td>Amiodarone</td>
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<tr>
<td>Talc granulomatosis</td>
</tr>
<tr>
<td>Siderosis</td>
</tr>
<tr>
<td>Tannosis</td>
</tr>
<tr>
<td>Coal worker’s pneumoconiosis</td>
</tr>
<tr>
<td>Sulfasalazine</td>
</tr>
<tr>
<td>Minocycline</td>
</tr>
</tbody>
</table>

### Diffuse Parenchymal Lung Diseases

- **DPLD of known cause**
  - Drugs induced
  - Radiation therapy
  - Collagen vascular diseases
  - Systemic diseases
  - Occupational exposures
- **Granulomatous diseases**
  - Hypersensitivity pneumonitis
  - Sarcoidosis
- **Other DPLD: cystic, congenital lung diseases**
- **Idiopathic Interstitial Pneumonias**
Interstitial Lung Disease

**Idiopathic Interstitial Pneumonias**
- Idiopathic pulmonary fibrosis
- Idiopathic nonspecific interstitial pneumonia
- Respiratory bronchiolitis–ILD
- Desquamative interstitial pneumonia
- Cryptogenic organizing pneumonia
- Acute interstitial pneumonia
- Rare idiopathic interstitial pneumonia
- Idiopathic lymphoid interstitial pneumonia
- Idiopathic pleuroparenchymal fibroelastosis
- Unclassifiable idiopathic interstitial pneumonias

**Diagnostic Approach to ILD**
- **Clinical**
  - Smoking history
  - Medications, other drug history and treatments
  - Hobbies, travel
  - Exposures
    - Occupational
      - Industrial, agricultural
    - Environmental
      - Pets, bird feathers/down bedding, hot tubs, contaminated water sources
  - Family history
  - Comorbid diseases
## Diagnostic Approach to ILD

<table>
<thead>
<tr>
<th>Physical Examination</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Crackles, dry or velcro</td>
</tr>
<tr>
<td>• Clubbing</td>
</tr>
<tr>
<td>• Signs of right heart strain/failure</td>
</tr>
<tr>
<td>• Signs of systemic disease (vasculitis, connective tissue diseases)</td>
</tr>
<tr>
<td>» Potential biopsy sites (rashes)</td>
</tr>
</tbody>
</table>

### Pulmonary Function Testing

- Interstitial inflammation and scarring results in restrictive defect
- Impaired gas exchange with a reduced diffusing capacity
- Measures of O2 saturation with exercise
  » 6 Minute walk
- Not diagnostic but characterizes severity

- Obstructive physiology not typical features of ILD
  » May be present with coexisting COPD
# Diagnostic Approach to ILD

## Laboratory Testing
- No specific laboratory tests or biomarkers
- Routine laboratory testing with chemistries, CBC with differential
- Evaluation for autoimmune diseases
  - ANA/ENA
  - Rheumatoid factor/CCP
  - CK, aldolase
  - If signs /symptoms of vasculitis, consider ANCA

## Chest imaging
- CXR findings nonspecific
- High resolution chest CT is central in the diagnosis and evaluation of ILDs
  - Patterns suggestive of certain disorders
  - Replaced biopsy in some cases

## Lung biopsy
## Case

- Diagnosed with interstitial lung disease and hypoxemia
- Referred to pulmonary
- Chest CT showed interstitial lung disease
- Lung biopsy with diagnosis of Usual Interstitial Pneumonitis (UIP)
- Idiopathic Pulmonary Fibrosis

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## Idiopathic Pulmonary Fibrosis

- Most common ILD of unknown etiology
- Mainly affects over age of 50, most over 60
- Incidence is estimated at 7.4-17 cases per 100,000 per year
- Prevalence of IPF is estimated at 13-60/100,000
- More men than women (1.5:1 ratio)
- 5-15% have a familial form
  - Present at a younger age
- Possible risk factors for developing IPF include cigarette smoking, occupational/environmental exposures (dusts)
What causes IPF?

<table>
<thead>
<tr>
<th>#1 Genetic Predisposition</th>
<th>#2 Epithelial Injury</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Surfactant proteins C</td>
<td>• Inhaled exposures</td>
</tr>
<tr>
<td>• Surfactant protein A2</td>
<td>• Dusty environment</td>
</tr>
<tr>
<td>• Telomerase genes</td>
<td>(organic and inorganic</td>
</tr>
<tr>
<td>• TERT, TERC</td>
<td>materials)</td>
</tr>
<tr>
<td>• 18% familial cases</td>
<td>• Tobacco smoke</td>
</tr>
<tr>
<td>• Mucin (MUC) 5B</td>
<td>• Viruses</td>
</tr>
<tr>
<td>• 1/3rd sporadic cases</td>
<td>• Acid reflux/aspiration</td>
</tr>
</tbody>
</table>

Familial Pulmonary Fibrosis

- Patients look just like IPF
- Typically ages 50-70
- Definition: first degree relative with IPF
- Probably autosomal dominant with variable penetrance
- Accounts for 5-15% of patients with IPF
- Genetic cause found in about 10% of familial pulmonary fibrosis
- Treatment is the same as IPF
<table>
<thead>
<tr>
<th>Idiopathic Pulmonary Fibrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>• History/Exam</strong></td>
</tr>
<tr>
<td>• Gradual onset and progressive dyspnea</td>
</tr>
<tr>
<td>• Nonproductive cough</td>
</tr>
<tr>
<td>• Bibasilar inspiratory crackles (Velcro crackles)</td>
</tr>
<tr>
<td>• Clubbing also common</td>
</tr>
<tr>
<td>• Later in the clinical course, signs of right heart failure, peripheral edema, cyanosis</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Idiopathic Pulmonary Fibrosis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>• PFTs show restriction, low diffusing capacity and desaturation with exertion</strong></td>
</tr>
<tr>
<td><strong>• +ANA, +RF unclear clinical significance</strong></td>
</tr>
<tr>
<td>• Diagnosis confirmed by imaging, lung biopsy</td>
</tr>
</tbody>
</table>
## Chest CT in IPF

- Subpleural basal predominance
- Reticular abnormality
- Honeycombing
- Traction bronchiectasis
- Absence of features listed as inconsistent with UIP pattern
  - Upper lung or mid lung predominance
  - Ground-glass abnormality, nodules, discrete cysts, mosaic attenuation/air trapping, consolidation
- Presence of these findings on HRCT in a patient without evidence of an alternative diagnosis
- Sufficient for a confident diagnosis of IPF
- Accuracy of 79-90%

## Normal Chest CT
Chest CT: subpleural reticular infiltrates

Chest CT: traction bronchiectasis
Chest CT: basilar honeycomb infiltrates

Role of Lung Biopsy

- In about 1/3rd of patients, require tissue to confirm diagnosis
  - Atypical findings on CT scan or clinical history
  - Early in disease process
Role of Lung Biopsy

• Bronchoscopy with transbronchial biopsy
  • Bronchoscopic biopsy does not confirm diagnosis of IPF
  • Useful to evaluate for alternate diagnosis
    » Granulomatous disorders (sarcoidosis, hypersensitivity pneumonitis)
    » Malignancy, lymphangitic carcinomatosis
    » Eosinophilic pneumonia, alveolar proteinosis, Langerhans
    » Bacterial, viral, and fungal infections
• Thoracoscopic lung biopsy (VATS)

IPF Lung Pathology: UIP

• Usual Interstitial Pneumonitis (UIP pattern)
• Evidence of marked fibrosis, architectural distortion
• Honeycombing in a predominantly subpleural/paraseptal distribution
• Presence of patchy involvement of lung parenchyma by fibrosis
• Presence of fibroblast foci
• Absence of features against a diagnosis of UIP suggesting an alternate diagnosis
### IPF Lung Pathology: UIP

<table>
<thead>
<tr>
<th>Fibrosis with collagen deposition</th>
<th>Temporal heterogeneity</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image1.png" alt="Image 1" /></td>
<td><img src="image2.png" alt="Image 2" /></td>
</tr>
</tbody>
</table>

### IPF Lung Pathology: UIP

<table>
<thead>
<tr>
<th>Fibroblastic foci</th>
<th>Microcystic changes</th>
</tr>
</thead>
<tbody>
<tr>
<td><img src="image3.png" alt="Image 3" /></td>
<td><img src="image4.png" alt="Image 4" /></td>
</tr>
</tbody>
</table>
Causes Of Usual Interstitial Pneumonitis

- Idiopathic pulmonary fibrosis (IPF)
- Collagen vascular disease
  - Rheumatoid arthritis
- Drug toxicity, radiation-induced
- Post-inflammatory pulmonary fibrosis
- Chronic hypersensitivity pneumonitis
  - May see granulomas or other clues of HP
- Occupational exposures
  - Asbestosis
- Familial idiopathic pulmonary fibrosis
- Hermansky–Pudlak syndrome

Clinical Course of IPF

- Unpredictable course for an individual patient
- Progressive disease
- Median survival of about 3-5 years
- Cause of death in about ½ related to IPF and respiratory failure
- Others: CAD/MI, infection, strokes
- Limited treatment options in the past
- Lung transplant
Coexisting conditions with IPF

- **Pulmonary hypertension**
  - In about 1/3 patients and most with advanced disease
  - Associated with worse pulmonary function, hypoxemia
  - Decreased exercise capacity and worse survival

- **GERD**
  - Common in IPF (65-94%)
  - Potential causal relationship between GERD and IPF through microaspiration of gastric contents
  - Acid-suppression therapy was associated with a slower rate of decline in pulmonary function and longer survival

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Coexisting conditions with IPF

- **Combined Pulmonary Fibrosis and Emphysema**
  - ~8% IPF cases, male, smoking history
  - Disproportionately low DLCO and gas exchange
  - Chest CT upper lobe emphysema, lower lobe fibrosis
  - High incidence of pulmonary hypertension, lung cancer and worse prognosis

- **Lung Cancer**
  - Increased risk in IPF patients, independent of other risks (smoking)

- **OSA, CAD, depression**
## Acute Exacerbation of IPF

- Acute deterioration with rapid, irreversible clinical decline
  - 1 and 3-year incidence of AE estimated 14% and 21%
  - Mortality rate associated with AE as high as 50% to 80%
  - Survival times 4-15 months in those who “recover”

- Etiology of decline unknown
- Chest imaging shows diffuse ground glass infiltrates
- Lung biopsy shows diffuse alveolar damage (identical to ARDS) superimposed on UIP pattern

## Acute Exacerbation of IPF

- Clinical evaluation to rule out an identifiable cause
  - Infection
    » Consider bronchoscopy
    » Often limited by hypoxemia and risk of respiratory failure
  - Progressive heart failure, ischemic disease
    » ROMI, Echo, BNP
  - Pulmonary embolism
    » CTPE study, LE duplex
- No well established therapy
IPF Diagnosis Requires
A Multi-Disciplinary Approach

Know your patient
Diseases, exposures, meds, family
** Pulmonary Fibrosis ≠
Idiopathic Pulmonary Fibrosis **

Get to know your radiologist

• Agree on definition of UIP
• Presence or absence of honeycombing
• Presence or absence of ground glass infiltrates
• Geographic location of infiltrates

**Diffuse interstitial infiltrates ≠ IPF**
**Get to know your thoracic surgeon**

- 2 or 3 lobe biopsies
- Avoid the tip of the lingula, middle lobe or lower lobes
- Target ground glass infiltrates or transition zones

**End stage fibrosis ≠ IPF**

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**Get to know your pathologist**

- Presence or absence of fibroblastic foci
- Temporal heterogeneity?
- Presence or absences of microcystic changes
- Presence or absence of collagen deposition
- Granulomas?

**End stage fibrosis ≠ IPF**
IPF Treatment: What Works?

Jim Allen, MD
Medical Director, University Hospital East
Professor of Internal Medicine
Division of Pulmonary & Critical Care Medicine
The Ohio State University Wexner Medical Center

IPF Treatment: What Works?

• Oxygen
• Pulmonary rehabilitation
• Lung transplant
• Esophageal reflux treatment

• Pirfenidone
• Nintedanib
• Sildenafil (?)
## Home oxygen options

<table>
<thead>
<tr>
<th>Stationary home units:</th>
<th>Portable units</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Oxygen concentrators</strong></td>
<td><strong>Compressed gas tanks</strong></td>
</tr>
<tr>
<td>– Standard (1-5 L)</td>
<td>– E cylinders</td>
</tr>
<tr>
<td>– High flow (10 L)</td>
<td>– M-6 cylinders</td>
</tr>
<tr>
<td><strong>Liquid oxygen reservoir</strong></td>
<td><strong>Portable liquid oxygen tanks</strong></td>
</tr>
<tr>
<td></td>
<td><strong>Portable oxygen concentrators</strong></td>
</tr>
</tbody>
</table>

## Pulmonary Rehabilitation

- 8 week programs
- 3 days per week
- 1-2 hours per session
- Focus on:
  - Education
  - Aerobic conditioning
  - Quality of life
Effect of pulmonary rehabilitation on interstitial lung disease


Pulmonary rehabilitation in IPF

- Significant improvement in 6MWT distance
- Significant improvement in fatigue severity scale

Swigris, et al. Respir Care 2011; 56:783-9
Pulmonary rehabilitation in interstitial lung disease


6MWT improvement after pulmonary rehabilitation for interstitial lung disease

Lung transplant contraindications

- Age > 65 (sort of…)
- BMI > 30
- Smoking in the past 6 months
- Uncured malignancy
- HIV, active hepatitis C/B
- Active infection
- Chest wall deformity
- Non-compliance
- Inadequate psychosocial support

Survival after lung transplant for IPF

Survival after lung transplant for IPF


Esophageal reflux and IPF mortality

- Treatment
- No treatment
### IPF Treatment: What Doesn’t Work?

- Corticosteroids
- Azathioprine
- Cyclophosphamide
- Everolimus
- Anticoagulation
- N-acetylcysteine
- Bosentan
- Ambrisentan
- Interferon-gamma
- Etanercept
- Imatinib
- Ribaviran

### New drugs for IPF

- Nintedanib*
- Pirfenidone*

*Approved by the FDA October 15, 2014*
Pirfenidone (ASCEND trial)

- Reduced loss of lung function (FVC)
- Reduced loss of exercise tolerance
- Improved progression-free survival

Pirfenidone versus Placebo

<table>
<thead>
<tr>
<th>Weeks</th>
<th>Change in FVC (ml)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>12</td>
<td>-50</td>
</tr>
<tr>
<td>24</td>
<td>-100</td>
</tr>
<tr>
<td>36</td>
<td>-150</td>
</tr>
<tr>
<td>52</td>
<td>-200</td>
</tr>
</tbody>
</table>

Legend:
- Pirfenidone
- Placebo
# Pirfenidone

### Dosing:
- **Week 1:** One capsule three times daily with food
- **Week 2:** Two capsules three times daily with food
- **After week 3:** Three capsules three times daily with food
- Dose can be reduced if side effects occur

### Laboratory monitoring:
- LFTs monthly x 6 months
- LFTs every 3 months thereafter
- **Dose adjustments:**
  - LFTs 3-5 times normal: reduce dose to 100 mg every 12 hours
  - LFTs > 5 times normal: stop pirfenidone

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# Nintedanib (IMPULSIS trials)

- Reduced loss of lung function (FVC)
- Reduced time to first exacerbation (IMPULSIS-2 trial)
**Nintedanib versus Placebo**

![Graph showing change in FVC (ml) over weeks for Nintedanib and Placebo](image)

**Dosing:**
- 150 mg every 12 hours with food
- Dose can be reduced to 100 mg every 12 hours if side effects occur

**Laboratory monitoring:**
- LFTs monthly x 3 months
- LFTs every 3 months thereafter
- Dose adjustments:
  - LFTs 3-5 times normal: reduce dose to 100 mg every 12 hours
  - LFTs > 5 times normal: stop nintedanib
## Side Effects:

<table>
<thead>
<tr>
<th></th>
<th>Pirfenidone</th>
<th>Nintedanib</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea</td>
<td>36%</td>
<td>24%</td>
</tr>
<tr>
<td>Rash*</td>
<td>28%</td>
<td></td>
</tr>
<tr>
<td>Adverse effect requiring discontinuation</td>
<td>14%</td>
<td>21%</td>
</tr>
</tbody>
</table>

*Photosensitivity

## Pirfenidone and Nintedanib: practical considerations

- Both drugs roughly equally effective
- Both drugs very expensive ($90-100,000/year)
- If patients are intolerant of one, consider changing to the other
- Giving both drugs together is **NOT** advised
Which patients benefit most from treatment?

- We don’t know
- Probably patients with earlier stage disease
  - FVC > 50% and DLCO > 30%
  - Patients with advanced disease are untested
- We do not know about non-IPF conditions:
  - Post-inflammatory pulmonary fibrosis
  - Rheumatoid arthritis-associated ILD
  - Chronic hypersensitivity pneumonitis

These drugs do not cure, they merely slow down the progression of the disease

Cure
Sildenafil Prevents Loss Of 6 MWT Distance In IPF Patients with RVH

Sildenafil in IPF*

- No nitrates or unstable angina
- Initial dose: 20 mg then monitor for 1 hour:
  - Symptoms
  - Blood pressure
  - Oxygen saturation
- Maintenance dose: 20 mg three times daily

Sildenafil is not FDA-approved for treatment of IPF
Typical Clinical Course

When patients with IPF are worse:

- Progression of IPF
- Anemia
- Heart failure
- Pulmonary embolism
- Lung cancer
- Infection
- Pneumothorax
“Stair-Step” Clinical Course

Disability

Death

Time (years)

Acute interstitial pneumonitis

- Diagnosis of exclusion
- Sudden-onset of worsened oxygenation and ground glass infiltrates
- Lung biopsy = diffuse alveolar damage (identical to ARDS)
- Steroids may help
Acute Interstitial Pneumonitis

April, 2013

August, 2014

Hyaline membranes
Ground glass infiltrates

Acute interstitial pneumonitis is a diagnosis of exclusion

- Heart failure
  - Consider BNP
  - Consider cardiac echo

- Pulmonary embolism
  - Consider CT-PA

- Infection
  - Consider BAL
Cough and IPF

Are there other causes:
- ACE inhibitors?
- Chronic rhinitis?
- Asthma/COPD?
- Reflux?

Palliating the IPF cough:
- Non-opioid anti-tussives (eg, benzonatate)
- Opioids
- Nebulized lidocaine?
- Thalidomide?
- Low dose corticosteroids?

Thalidomide Reduces Cough In IPF

<table>
<thead>
<tr>
<th>Cough Quality of Life Questionnaire</th>
<th>Cough Visual Analogue Scale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baseline</td>
<td>Baseline</td>
</tr>
<tr>
<td>Placebo</td>
<td>Placebo</td>
</tr>
<tr>
<td>Thalidomide</td>
<td>Thalidomide</td>
</tr>
</tbody>
</table>

Fatigue and IPF

- Anemia?
- Thyroid disease?
- Sleep apnea?
- Heart failure?
- Exertional hypoxemia?

Sleep apnea is common in IPF:

- Incidence* = 88%!!!
  - 20% mild
  - 68% moderate-severe
- Undiagnosed sleep apnea contributes to fatigue
- Quality of life can improve with CPAP

* Chest 2009; 136:772-778
What else can you do to improve the quality of life?

- Smoking cessation
- Maintenance of a normal BMI
- Vaccinations
- Recognize and treat depression
Vaccinations for patients with IPF:

- Influenza
- Pertussis (Tdap)
- Strep pneumoniae

New CDC Pneumococcal Vaccine Recommendations:

- Adults < 65 and low risk: vaccine not required
- Adults < 65 and moderate risk
  - PPSV-23
- Adults < 65 and high risk
  - PCV-13
  - PPSV-23 6-12 months later
  - Repeat PPSV-23 in 5 years
- Adults > 65
  - PCV-13
  - PPSV-23 6-12 months later
  - Repeat PPSV-23 in 5 years
Idiopathic pulmonary fibrosis is ultimately a terminal disease

Start end-of-life discussions early

- Resuscitation and intubation
- Hospice
- How patients die

Photo: Anthony Majanlathi
Galata Morente, Capitoline Museum, Rome
Outcome of patients admitted to the ICU with respiratory failure

- Die in ICU
- Die in hospital
- Discharged home

Hospice

- Anticipated life expectancy < 6 months
- Levels of care:
  - Routine home care
  - Continuous home care
  - Inpatient care
  - Respite care

- Physician services
- Nursing services
- Social services
- Supplies
- Medications
- Bereavement counseling
- Hospice aide
- PT/OT/ST
How do patients with IPF die?

The improvement in the outcomes of your patients tomorrow will depend on clinical trials in your patients today.
IPF Treatment: Summary

• Establish a confident diagnosis!
• New drugs: nintedanib & pirfenidone
• Don’t do things that don’t work
• Consider clinical trials
• Never miss an opportunity for transplant
• The little things make a big difference in quality of life

Case #1
Case #1

Case #2