Idiopathic Pulmonary Fibrosis

The main thing you have to do when considering IPF is to rule out other causes of interstitial lung disease.

<table>
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<th>History</th>
<th>Physical Exam</th>
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<td>50-60 years old</td>
<td>Coarse bibasilar crackles at end inspiration</td>
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<td>Most have a smoking history</td>
<td>• Can be unilateral especially early</td>
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<td>Several months of dyspnea and cough</td>
<td>End inspiratory squeaks are traction bronchiectasis</td>
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<tr>
<td>Systemic symptoms are rare (fever, myalgias, etc)</td>
<td>Clubbing in 50-75%</td>
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**History**

**Physical Exam**

- Coarse bibasilar crackles at end inspiration
- End inspiratory squeaks are traction bronchiectasis
- Clubbing in 50-75%

**Radiology**

If the H&P is as above and the CT chest has these three features, then it is likely IPF

1. Peripheral, bibasilar predominant reticular opacities
2. Honeycombing
3. Traction bronchiectasis

However, there are a few exceptions to the above— as experts you should be aware:

- Honeycombing is essential to making a UIP diagnosis, but is sometimes absent
- Up to 30% of UIP diagnosed by biopsy have other features on CT chest than listed above
- Ground glass opacities can be present in IPF! But should not be the predominant feature

**So, if I’m pretty sure it’s IPF, do I need labs?** Usually the answer is no. See the ILD handout for details on serologies. A few things to keep in mind:

1. ANA >1:40 is present in up to 25% of biopsy proven UIP
2. Positive rheumatoid factor is present in about 15%
3. Up to 70% a people with anti-synthetase syndrome can present with a UIP pattern of fibrosis

**When do I recommend a biopsy?**

If with the above, you are not confident in an IPF diagnosis then get a biopsy. It’s important to get this right because IPF has a prognosis worse than most cancers. But usually you can spare them the morbidity of a biopsy.

**Ok. So they have IPF. What can I tell them to expect?**

- FVC declines about 150 – 200 cc per year – this means they can expect a slow decline in functioning
- Acute exacerbations are unpredictable – this means consider early transplant referral
- “If I take 100 people with IPF, about 20 of them will be dead in 5 years”

And then they say, “aren’t there some medicines or something?” For mild to moderate IPF, meaning DLCO 45-65% predicted, I would have a discussion about the pros and cons of medication. Here’s what you need to know:

- **Nintedanib** is a multiple tyrosine kinase blocker and mediates elaboration of fibrogenic growth factors (PDGF, VEGF, FGF). It slows the rate of decline in lung function. Don’t give to cirrhotics. And it interferes with CYP3A4 and increases risk of bleeding on anticoagulation.
- **Pirfenidone** is an antifibrotic that inhibits TGF-beta stimulated collagen synthesis; blocks fibroblast proliferation in vitro. Also slows the rate of decline in lung function and possibly a survival benefit in a pooled analysis.

Now they are in the hospital with an acute exacerbation of IPF. So now what? Know that mortality is about 50% and probably higher if they end up on mechanical ventilation – approaching 100% in some studies. Can try the following:

- Rule out infection and give 1-2 grams of solumedrol daily for 3-5 days- know that evidence for this is lacking
- Very limited data on protocol with pheresis and IVIg- patients that respond to this (and the above meds) probably have an underlying, undiagnosed connective tissue disease