Interstitial Lung Disease on the MAU
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ILD – The Confusing Land of the TLA?

• Take it back to basics
  – Deliberately avoiding lots of science

• Discuss the challenges of clinical practice
  – Diagnosis
  – AE-ILD
  – Palliative Care
  – EOLC

• Questions
Interstitium
Fibrosis (Pink)
Inflammation (Blue)
ILDs
>200

Known Cause
eg. CTD/Drugs/Asbestos

Idiopathic Interstitial Pneumonias

Granulomatous Disease
eg. Sarcoid and Hypersensitivity Pneumonitis

Others
eg. LAM/LCH

Idiopathic Pulmonary Fibrosis

Others
DIP RBILD NSIP AIP LIP COP
ILDs

>200

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e.g. CTD/Drugs/Asbestos

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e.g. Sarcoid and Hypersensitivity Pneumonitis

Others
e.g. LAM/LCH

Idiopathic Pulmonary Fibrosis

Others

DIP RBILD

NSIP AIP

LIP COP
Idiopathic Pulmonary Fibrosis (IPF)

One in a hundred people in the UK will die of IPF
Natural History of IPF

Think of ILD as a possible diagnosis
Crep = French Pancake ≠ Crackle
Lung Function

This is NOT COPD

<table>
<thead>
<tr>
<th>SPIROMETRY</th>
<th>MEASURED</th>
<th>PREDICTED</th>
<th>% PREDICTED</th>
<th>Z SCORE</th>
</tr>
</thead>
<tbody>
<tr>
<td>FEV1 (l)</td>
<td>0.75</td>
<td>2.23</td>
<td>34</td>
<td>-3.90</td>
</tr>
<tr>
<td>VC (l)</td>
<td>0.90</td>
<td>2.65</td>
<td>34</td>
<td>-4.06</td>
</tr>
<tr>
<td>FEV1/VC</td>
<td>83</td>
<td>84</td>
<td>99</td>
<td>-0.15</td>
</tr>
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</table>
Protocolised HRCT

1mm helix done prone with sharpened protocol
In both full inspiration and expiration
No contrast
Treatment of IPF
Steroids in IPF

Now the drugs don't work
They just make you worse
But I know I'll see your face again......

The Verve 1997
Anti-fibrotic Therapies (If MDT ratified IPF and VC 50-80%)

- Pirfenidone
- Nintedanib
Pirfenidone

• TGF-β inhibition

• Side effects: GI upset, weight loss, photosensitive rash, hepatotoxicity

• C/I: renal impairment eGFR <30
Pirfenidone rash
Nintedanib

• Triple TKI

• Side effects: GI upset, diarrhoea, hepatotoxicity

• Cautions: anticoagulants, cardiac disease, surgery, liver disease
Nintedanib Diarrhoea
Practical Point #2

Anti-fibrotic drugs are not immunosuppressive and don’t need to be stopped with infection
There is no cure for IPF

**Diagram Description**

- **Onset of Disease**
- **Onset of Symptoms**
- **Diagnosis**
- **Sub-clinical Period**
- **Pre-diagnosis Period**
- **Post-diagnosis Period**

**Time**

- 1 yr
- 2 yr
- 3 yr
- 4 yr
- 5 yr
- 6 yr

Integrated Palliative Care

Integrated Palliative Care
Co-Morbidities

- Emphysema
- Lung Cancer
- Pulmonary hypertension
- Venous thromboembolism
- Sleep-disordered breathing
- Gastroesophageal reflux disease
- Coronary artery disease
- Depression and anxiety
- Deconditioning
- Frailty and Sarcopenia
Combined Pulmonary Fibrosis and Emphysema (CPFE)

- Spirometry can be normal precluding antifibrotics
- Gas transfer very low / desaturate++
- Pulmonary hypertension common
- Poor prognosis
Lung cancer

- Increased risk in IPF (7x)
- Independent of smoking

- Treatment can trigger AE-IPF
Acute Exacerbations of IPF (AE-IPF)
AE-IPF

- 1 in 7 patients/year
- 20-40% fatality
- Sick enough to die
AE-IPF

Collard, H et al
Am J Respir Crit Care Med 2016 ; 194 :3, pp 265–275
AE-IPF

- Exclude PE
- Exclude heart failure
- Exclude infection

- Support with oxygen

- Steroid
- Antibiotics
- Co-trimoxazole 960mg bd

- Palliative care
- EOLC
- Do not (routinely) intubate and ventilate
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Lung transplantation?
Practice Point #3

Advanced care planning and guidance from the ILD team is critical...............if its not there then please ask for it!
Drug related ILD

www.pneumotox.com

The Drug-Induced Respiratory Disease Website
Philippe Camus, M.D.
Dijon, France

Patterns
- Pneumonitis (ILD), acute, severe (See also under ARDS)
- Pneumonitis (ILD), subacute, moderate
- Organizing pneumonia (OP/BOOP) - A pattern consistent with OP on imaging
- Pulmonary fibrosis
- Subclinical pulmonary opacities
- Lung nodule or nodules
- Diffuse alveolar damage (DAD) (see also under llb and XVI)
- ILD with a granulomatous component
- Nodules in lung parenchyma (a.k.a. nodulosis)
- Subclinical changes in lung function/PPFT
- Rapidly progressive ILD or pulmonary fibrosis (Hamman-Rich syndrome)
Immunosuppression?

NSIP (Often SSc)
- Ground glass predominant
- Peripheral, subpleural
- Basal predominant
- No honeycombing

UIP (Often RA)
- Little ground glass
- Peripheral, subpleural
- Basal predominant
- Honeycombing - key feature
Anti-synthetase ILD

- High index of suspicion
- Requires specific antibody testing
- Work closely with Rheumatology
Hypersensitivity Pneumonitis
Look Carefully
Look Carefully
Listen Carefully

The inspiratory “squawk” in extrinsic allergic alveolitis and other pulmonary fibroses

JE EARIS, K MARSH, MG PEARSON, CM OGILVIE

From the Cardiothoracic Centre, Broadgreen Hospital, Liverpool

Fig 3  Phonopneumogram and simultaneous inspiratory flow trace of a patient with extrinsic allergic alveolitis due to exposure to a budgerigar; a crackle immediately precedes the “squawk.”
HP Treatment

Remove antigen exposure

Immunosuppression
Practice Point #4

If it is an index presentation and/or there is a good chance of a non-IPF diagnosis then seek early Respiratory and Critical Care input...........

.........some disease presentations do well and should be supported on critical care............
It can be challenging.......
In Conclusion

1. Think of ILD as a diagnosis, take a history and listen carefully

2. It can be challenging.....MDT working is vital

3. One in a hundred of us will die of IPF

4. We all need to get better at integrating palliative care and talking about death
Questions.....?

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