Diffuse Lung Disease:  
A Survival Guide  
For The Non-Thoracic Radiologist

Norwegian Society of Thoracic Imaging  
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Sujal R Desai  
King's College Hospital, London
Background

- Diffuse lung diseases (DILDs) are an important but difficult group of disorders.
- HRCT is an important component of clinical investigation in patients with suspected DILD.
- HRCT interpretation *not* straightforward...
Problems...

- >200 recognised diffuse interstitial lung diseases
- ~ 10 pathological patterns
- ~ 5-6 HRCT manifestations
- 1 diagnosis
The Diffuse Pulmonary Lung Diseases: Classification

Diffuse Interstitial Lung Diseases

DILD of known cause
Drugs, collagen vascular disease

Granulomatous DILD
Sarcoidosis

"Others"
Langerhans’ cell histiocytosis, LAM

Idiopathic Interstitial Pneumonias
UIP, NSIP, DIP, RBILD, COP, AIP

ATS/ERS International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias *Am J Respir Crit Care Med* 2002;165:277
SURVIVAL GUIDE

Rule 1:

AVOID “OVERCALLING”
Interpretive Problems

“Overcalling”

The Influence of Expiratory Effort
Interpretive Problems

“Overcalling”

The Influence of Body Habitus
Interpretive Problems
“Overcalling”

The Influence of Age

Age 1

Age 10
Interpretive Problems

“Overcalling”

The Influence of Collimation

SURVIVAL GUIDE

Rule 2:

BE AWARE OF OBSERVER VARIABILITY
Interpretive Problems
Observer Variability

“...the appearances are of a interstitial lung disease and consistent with EXTRINSIC ALLERGIC ALVEOLITIS.”

(I haven’t got a clue...!)

- Observer Experience
- Atypical Radiological Appearances
- Non-specific CT features
Interpretive Problems
Observer Variability
SURVIVAL GUIDE
Rule 3:

KNOW THE COMMON HRCT SIGNS & PATHOLOGICAL CORRELATES
Common HRCT Patterns

Histopathological Correlates & Diagnoses

CONSOLIDATION ↔ AIR SPACE DISEASE

Infection

Tumour
- Adenocarcinoma
- Bronchoalveolar Cell Carcinoma
- 1º Pulmonary Lymphoma

Organizing Pneumonia

Wegener’s Granulomatosis

Eosinophilic Pneumonia

Sarcoidosis

Lipoid Pneumonia

etc...
Common HRCT Patterns
Histopathological Correlates & Diagnoses

RETICULAR PATTERN

INTERSTITIAL DISEASE

- CFA / IPF...other IIPs
- Asbestosis / Pneumoconioses
- Sarcoidosis
- Post-infective (eg TB)
- Lymphangitis
- Systemic sclerosis
- Berylliosis
- Drug-induced lung disease
  etc...
Common HRCT Patterns
Histopathological Correlates & Diagnoses

GROUND-GLASS OPACIFICATION

INTERSTITIAL DISEASE

AIR SPACE DISEASE

MIXED AIR SPACE & INTERSTITIAL DISEASE

Pulm Oedema
GROUND-GLASS OPACIFICATION

...a hazy increased opacity of lung, with preservation of bronchial and vascular margins. It is caused by partial filling of air spaces, interstitial thickening (due to fluid, cells, and/or fibrosis), partial collapse of alveoli, increased capillary blood volume or a combination of these, the common factor being the partial displacement of air.

Ground-Glass Opacification
The Physics…

**GROUND-GLASS OPACIFICATION**
...the common factor being the partial displacement of air

1.5mm
Ground-Glass Opacification
The Physics...
Ground-Glass Opacification
The Physics...
Ground-Glass Opacification
Dispelling a Few Myths...

CT CHEST:
...in addition, there is widespread ground-glass opacification bilaterally. The changes are most pronounced in the mid and lower zones.

IMPRESSION:
The appearances are those of a reversible (treatable) alveolitis...
Ground-Glass Opacification

Ground-Glass & Reversibility... Look at the Airways!
Ground-Glass Opacification
Tractional Dilatation of Airways

FROM: Google Images
SURVIVAL GUIDE

Rule 4:

TRY NOT TO “PLEASE” THE CLINICIAN!
Breathlessness and cough ... I suspect diffuse interstitial lung disease?

There is a subtle subpleural reticular pattern indicating “early” interstitial fibrosis...
SURVIVAL GUIDE

Rule 5:

KNOW THE COMMON / IMPORTANT DILDS
<table>
<thead>
<tr>
<th>HRCT diagnoses</th>
<th>Degree of Diagnostic Confidence / Accuracy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Idiopathic pulmonary fibrosis / CFA</td>
<td>++++/-</td>
</tr>
<tr>
<td>Langerhans’ cell histiocytosis</td>
<td>+++/-</td>
</tr>
<tr>
<td>Asbestosis</td>
<td>+++/-</td>
</tr>
<tr>
<td>Sarcoidosis</td>
<td>+++/-</td>
</tr>
<tr>
<td>Extrinsic allergic alveolitis</td>
<td>++/-</td>
</tr>
<tr>
<td>Alveolar proteinosis</td>
<td>++/-</td>
</tr>
<tr>
<td>(Cryptogenic) organizing pneumonia</td>
<td>+/-</td>
</tr>
<tr>
<td>Non-specific interstitial pneumonia</td>
<td>+/-</td>
</tr>
<tr>
<td>Smoking-related ILD</td>
<td>+/-</td>
</tr>
</tbody>
</table>
Idiopathic Pulmonary Fibrosis
Idiopathic Pulmonary Fibrosis

Key Background Facts

• Commonest of the idiopathic interstitial pneumonias

• Associated with histopathological/HRCT pattern of usual interstitial pneumonia (UIP) by definition

*NB Radiologist/pathologist makes diagnosis of pattern ... *not* synonymous with CFA/IPF

ATS/ERS International Multidisciplinary Consensus Classification of the Idiopathic Interstitial Pneumonias

*Am J Respir Crit Care Med* 2002;165:277
Idiopathic Pulmonary Fibrosis

Pathological Features

USUAL INTERSTITIAL PNEUMONIA

Peripheral (subpleural) predilection

Fibrosis with honeycombing

Temporal and Spatial Heterogeneity

IMAGES FROM:
Mueller-Mang et al. What every radiologist should know about idiopathic interstitial pneumonias Radiographics 2007;27:595-615
Idiopathic Pulmonary Fibrosis

Pathological Features

**USUAL INTERSTITIAL PNEUMONIA**

- Fibrosis with honeycombing
- Architectural destruction
- Peripheral and basal distribution
- Patchy (i.e. normal and abnormal lung)
- Fibroblastic foci

**IMAGES FROM:**
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Idiopathic Pulmonary Fibrosis
Characteristic HRCT Features

Reticular pattern with honeycombing
Idiopathic Pulmonary Fibrosis

Characteristic HRCT Features

UPPER ZONES vs LOWER ZONES
Idiopathic Pulmonary Fibrosis

Characteristic HRCT Features

- Reticular pattern *with* honeycombing
- Basal
- Subpleural

*High (>95%) positive predictive value of an (experienced) confident CT diagnosis* ¹ ²

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Idiopathic Pulmonary Fibrosis

Atypical HRCT Features

Langerhans’ Cell Histiocytosis
Langerhans’ Cell Histiocytosis

Key Background Facts

- Disease caused by infiltration by characteristic non-clonal antigen-presenting Langerhans’ cell – found in epithelial surfaces (e.g. epidermis, airways)

- LCH confined to lungs in >85%

- Pulmonary LCH strongly linked to cigarette-smoking
Langerhans’ Cell Histiocytosis

Histopathological Changes
Langerhans’ Cell Histiocytosis

Serial HRCT Changes
Langerhans’ Cell Histiocytosis

“Special” Features
Langerhans’ Cell Histiocytosis

Other Causes of “Cystic” Lung Disease

- (Bronchiectasis)
- Emphysema

- Lymphocytic interstitial pneumonia
- Extrinsic allergic alveolitis

- Lymphangioleiomyomatosis
- Tuberose sclerosis
- Neurofibromatosis

- Post-infective pneumatocele
Sarcoidosis
Sarcoidosis

Essential Pathology

- Mononuclear alveolitis
- Early, loosely formed granulomas
- Along lymphatic pathways
- Tightly packed N-C granulomas
  epithelioid histiocytes, multinucleate giant
  cells, surrounded by lymphocytes, monocytes and fibroblasts

Sarcoidosis

Pathological vs Radiological Features


Nishimura et al. Pulmonary Sarcoidosis: correlation of CT and histopathologic findings. Radiology 1993; 189:105-109
Sarcoidosis

HRCT Features
Sarcoidosis

Unusual Patterns

“Air Space”  
Asymmetrical

Mosaicism  
Thickened ILS

CFA look-alike
Extrinsic Allergic Alveolitis
Extrinsic Allergic Alevolitis
Radiological-Pathological Relationships
Extrinsic Allergic Alevolitis

Subacute Disease
Extrinsic Allergic Alevolitis

Chronic Disease ... “Head Cheese” Sign
Alveolar Proteinosis
Alveolar Proteinosis

The “Crazy Paving” Sign

IMAGE COURTESY: Dr Susan J Copley, Hammersmith Hospital, London

Alveolar Proteinosis

The “Crazy Paving” Sign

CAUSES

INFECTION

INFECTION
Pneumocystis carinii pneumonia (PCP)
Mucinous Bronchioloalveolar Carcinoma (BAC)

NEOPLASM

Pulmonary Alveolar Proteinosis (PAP)
Sarcoidosis
Nonspecific Interstitial Pneumonia (NSIP)
Organizing Pneumonia (OP)

IDIOPATHIC

Lipoid Pneumonia

INHALATION

Adult respiratory distress syndrome (ARDS)
Pulmonary Hemorrhage Syndromes

SANGUINEOUS

FROM: Rossi S et al. Crazy paving pattern at thin-section CT of the lungs: radiologic pathologic overview. Radiographics 2003; 23: 1509
Conclusions
Surviving the Minefield of DILDs

- Don’t overcall
- Be aware of “acceptable” observer variability
- Know the common HRCT patterns and their histopathological correlates
- Review clinical details AFTER reviewing the imaging
- Know the common DILDs and their HRCT features
An Invitation in 2012

20th Annual Meeting
22-24 June 2012 | Church House, London

European Society of Thoracic Imaging

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Pulmonary Vascular Disease • Infections in immune Compromise
Cardiac Imaging • Radiofrequency Ablation • Microwave Ablation
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