

Norwegian Society of Thoracic Imaging
Oslo, October 2011

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

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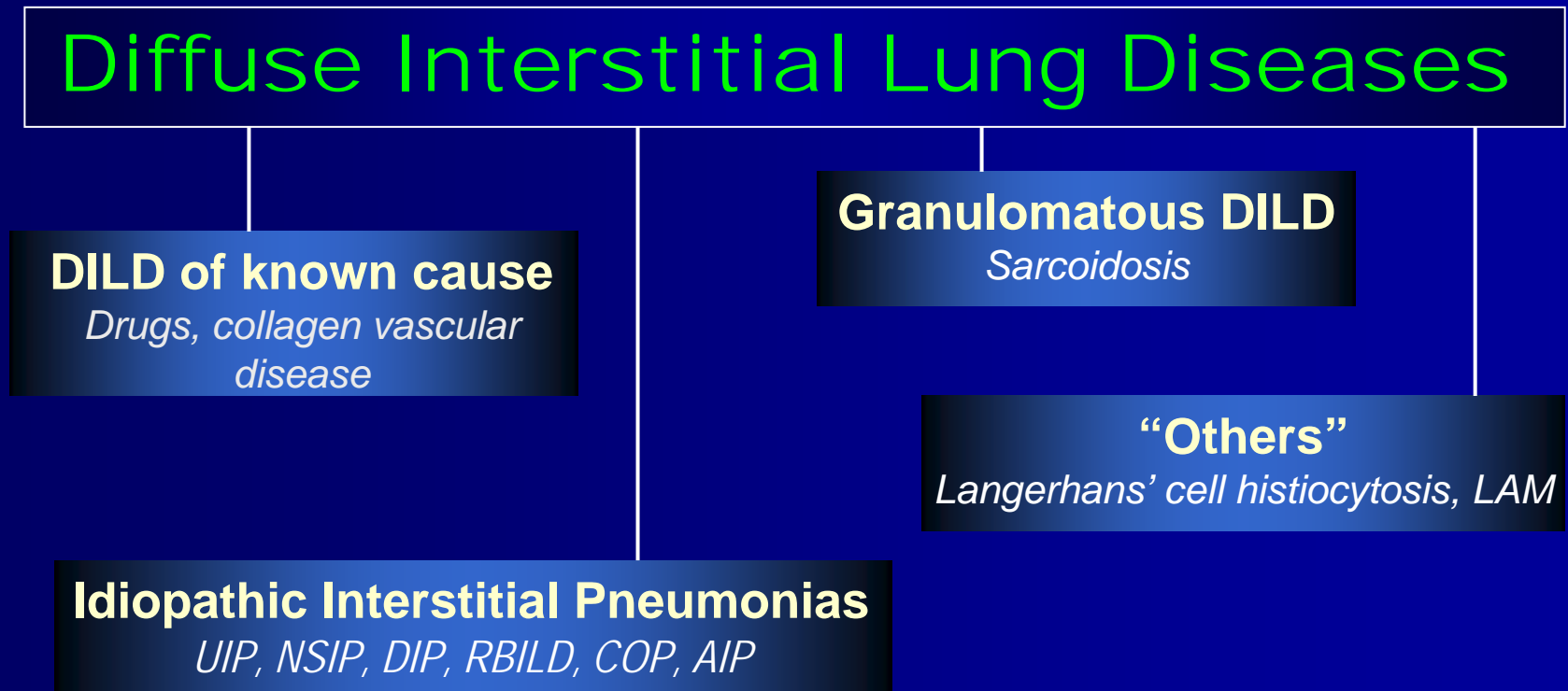
Background

- Diffuse lung diseases (DILDs) are a 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*



SURVIVAL GUIDE

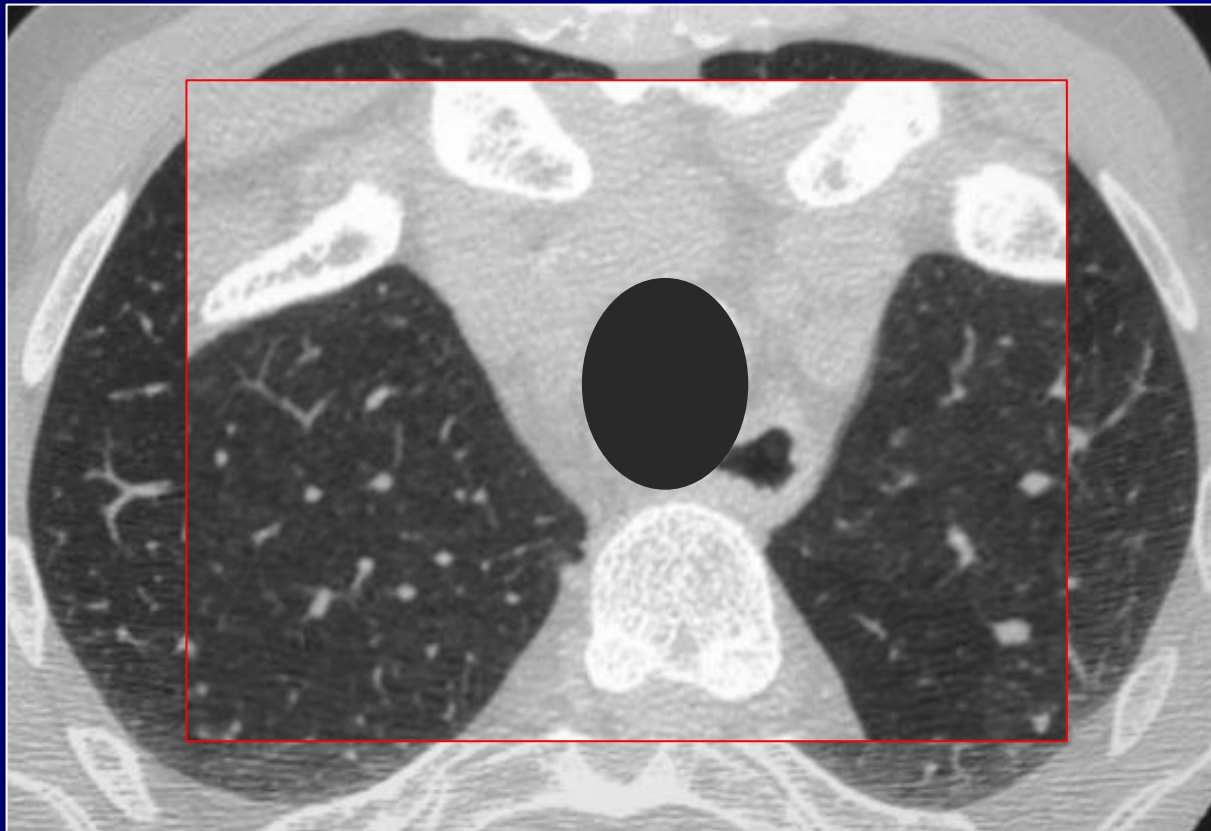
Rule 1:

AVOID “OVERCALLING”

Interpretive Problems

"Overcalling"

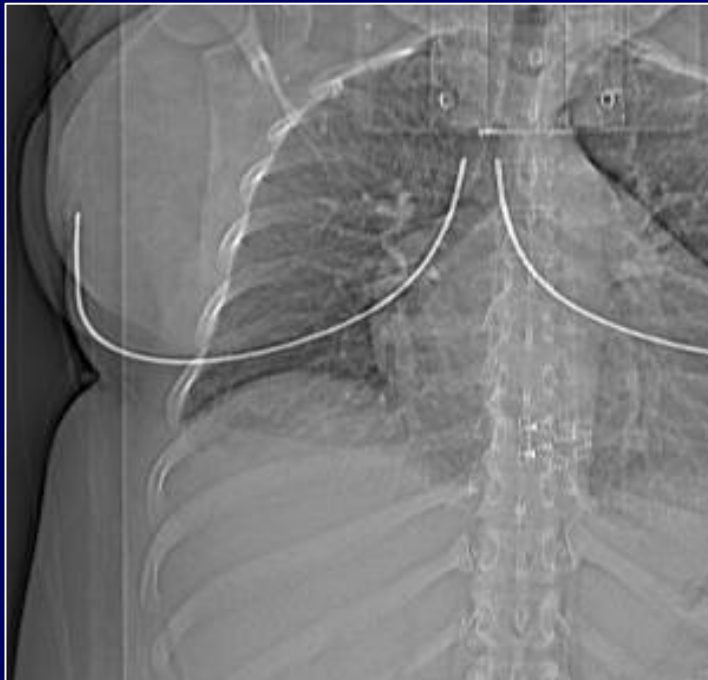
The Influence of Expiratory Effort



Interpretive Problems

"Overcalling"

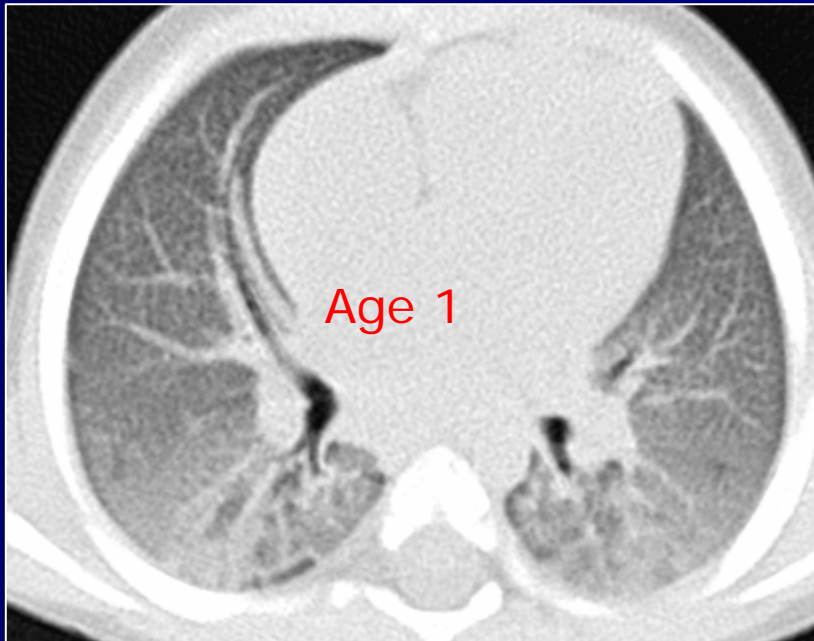
The Influence of Body Habitus



Interpretive Problems

"Overcalling"

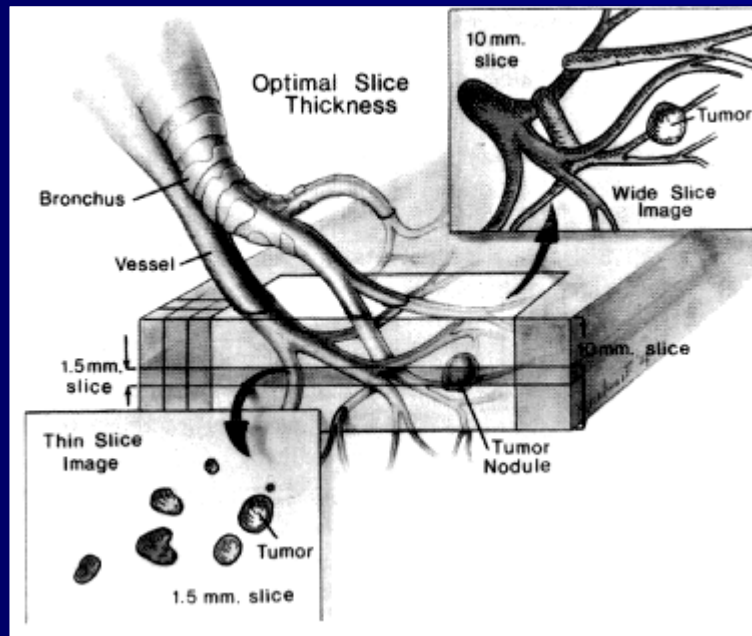
The Influence of Age



Interpretive Problems

"Overcalling"

The Influence of



FROM: Principles and Techniques of Thoracic Computed Tomography and Magnetic Resonance In: *Computed Tomography and Magnetic Resonance of the Thorax*. Naidich DP, Webb WR, Müller NL, Krinsky GA, Zerhouni EA, Siegelman SS (eds). 3rd Edition. Lippincott-Raven, Philadelphia

SURVIVAL GUIDE

Rule 2:

**BE AWARE OF
OBSERVER VARIABILITY**

Interpretive Problems

Observer Variability

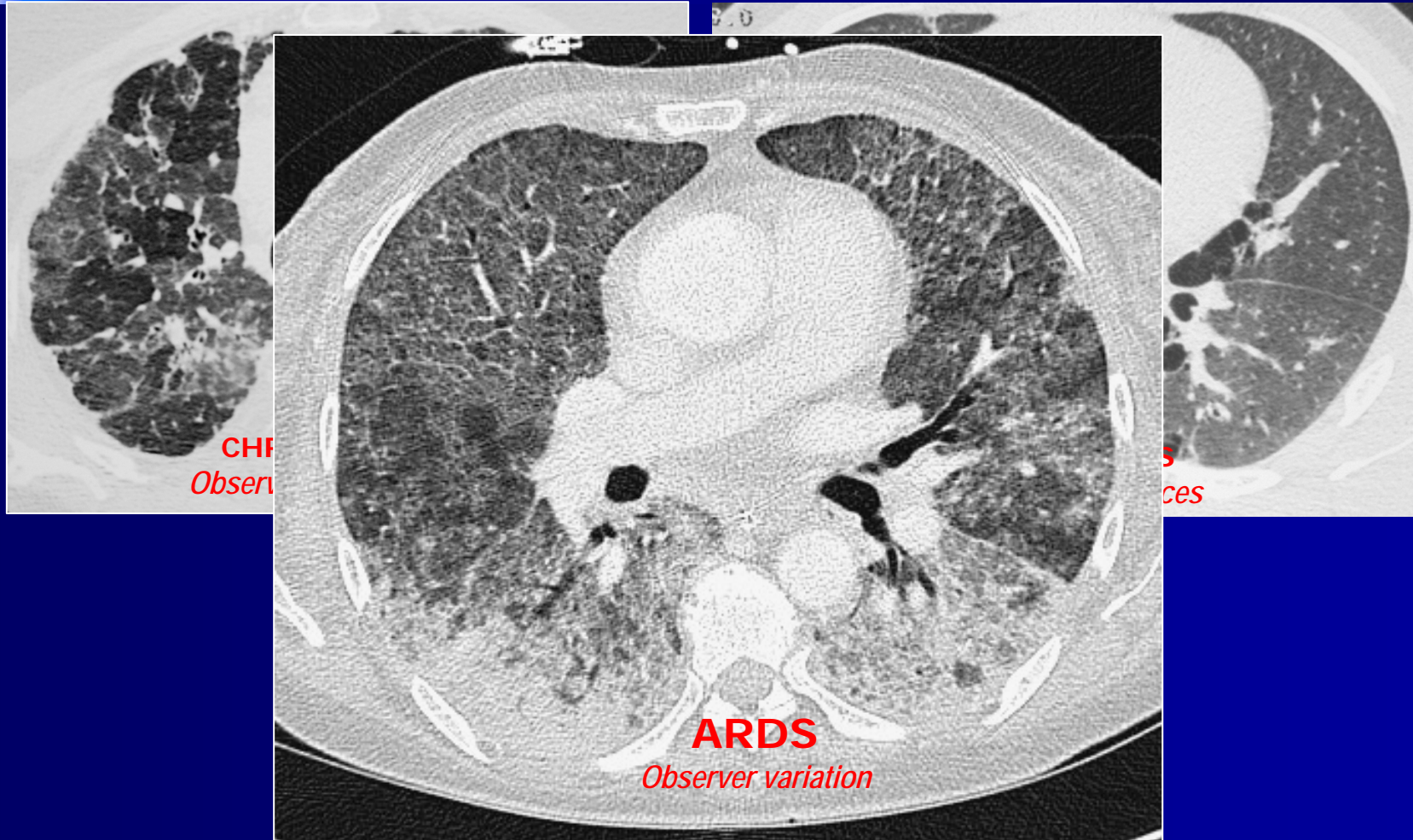
"...the appearances are of a interstitial lung disease and consistent with ~~EXTRINSIC ALVEOLAR PROTEINOSIS~~ **EXTRINSIC ALVEOLAR PROTEINOSIS**."

(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

1° Pulmonary Lymphoma

Organizing Pneumonia

Wegener's Granulomatosis

Eosinophilic Pneumonia

Sarcoidosis

Lipoid Pneumonia

etc...

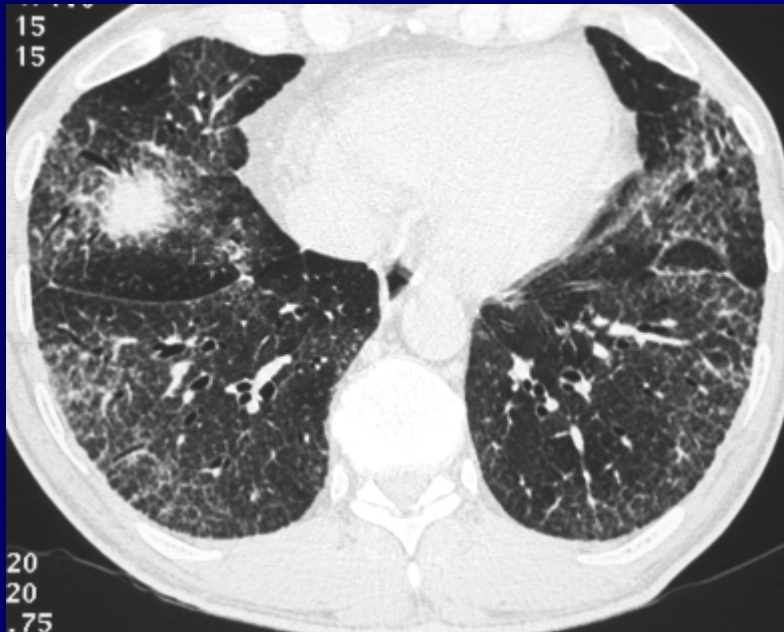
Common HRCT Patterns

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...

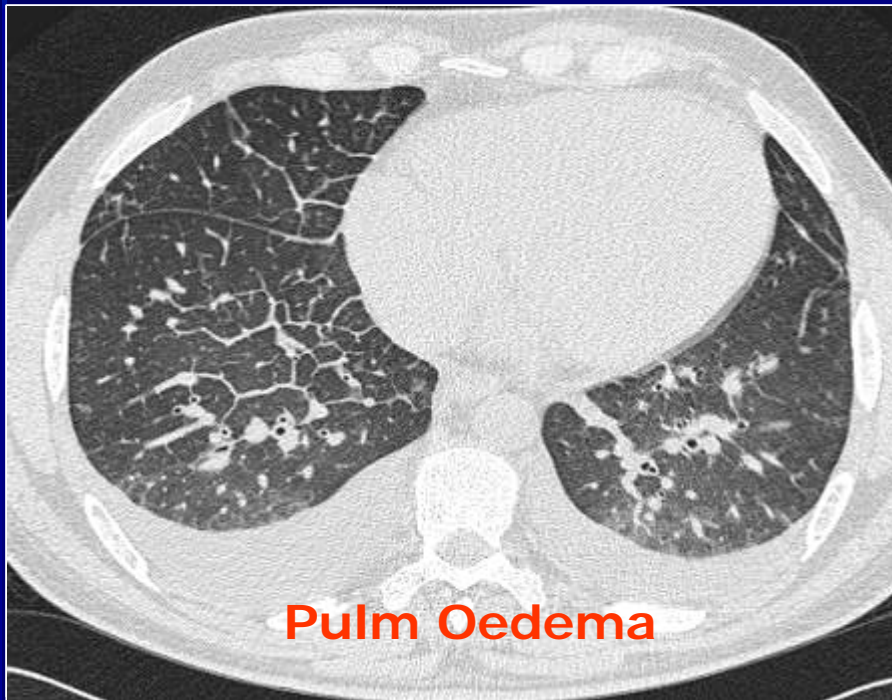
Common HRCT Patterns

Histopathological Correlates & Diagnoses

**GROUND-GLASS
OPACIFICATION**



INTERSTITIAL DISEASE

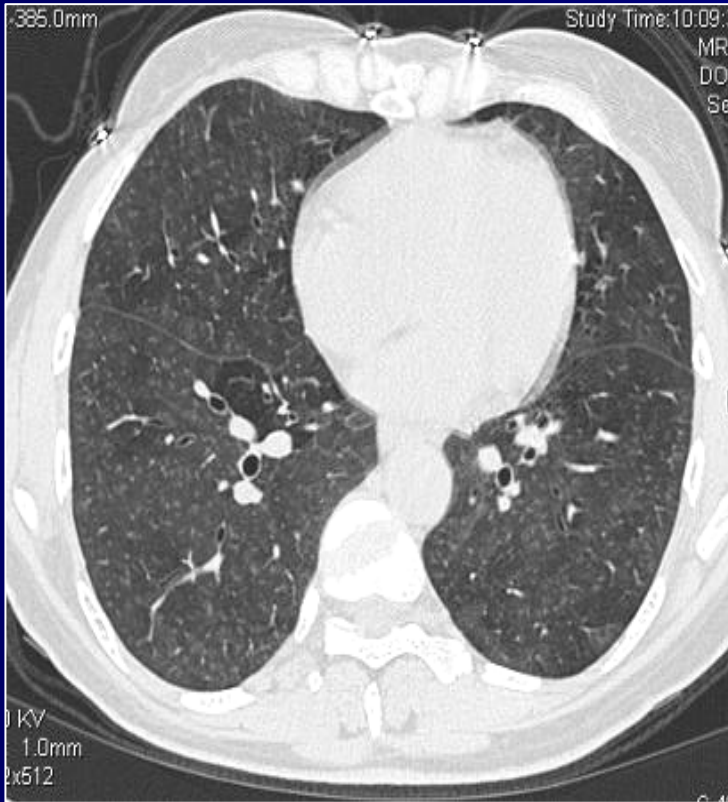


Pulm Oedema

AIR SPACE DISEASE

**MIXED
AIR SPACE
&
INTERSTITIAL
DISEASE**

Some definitions...



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

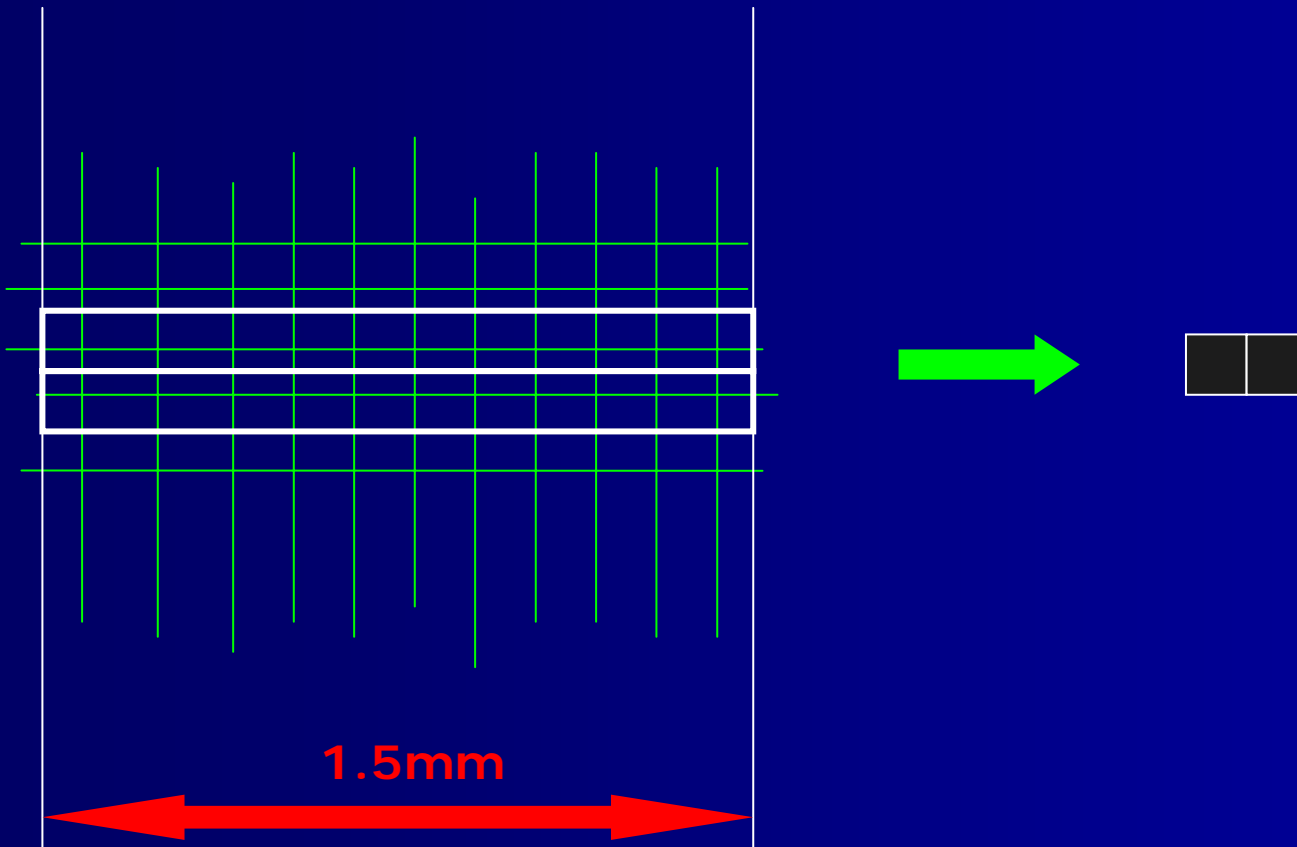
Hansell DM et al. Fleischner society: glossary of terms for thoracic imaging. *Radiology* 2008; 246:697-722

Ground-Glass Opacification

The Physics...

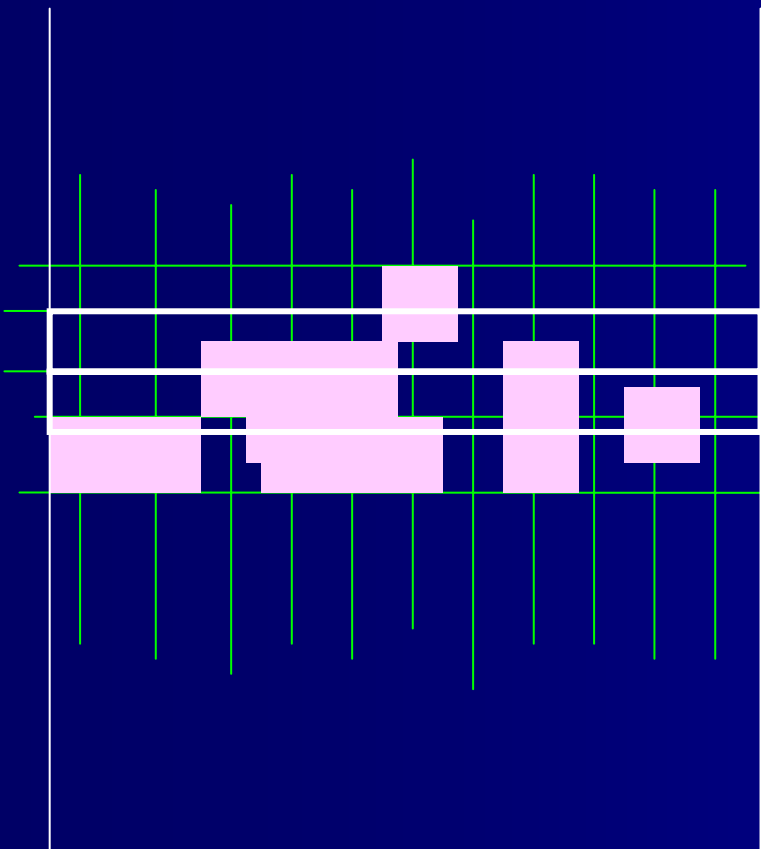
GROUND-GLASS OPACIFICATION

...the common factor being the partial displacement of air



Ground-Glass Opacification

The Physics...

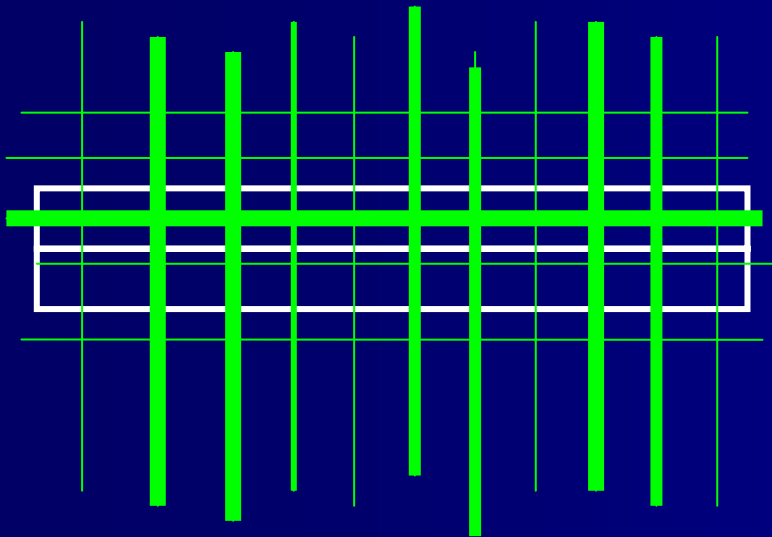


AIR↓



Ground-Glass Opacification

The Physics...



AIR↓



Ground-Glass Opacification

Dispelling a Few Myths...

CT CHEST:

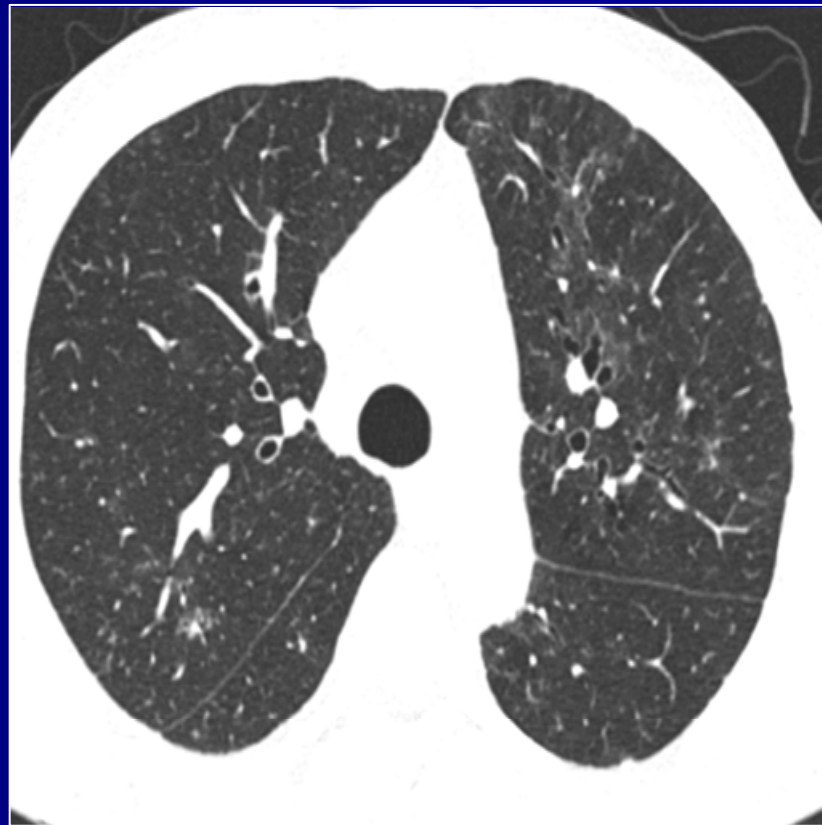
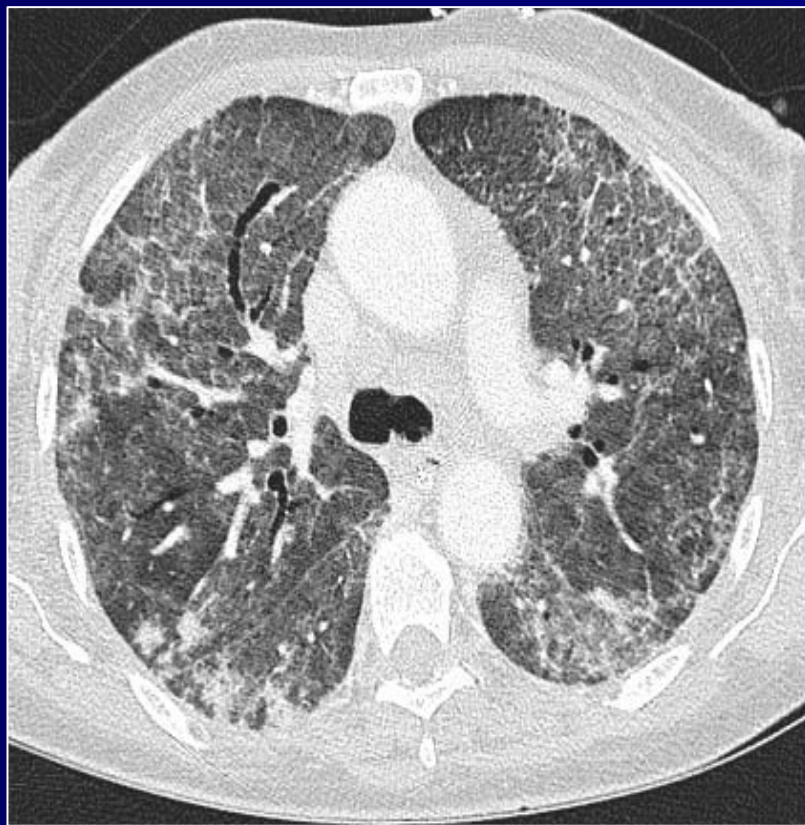
...in addition, there is widespread ground-glass opacification bilaterally. The changes are most pronounced in the mid and lower zones.

IMPRESSION:

The appearances are those of a reversible (treatable) alveolitis...

Ground-Glass Opacification

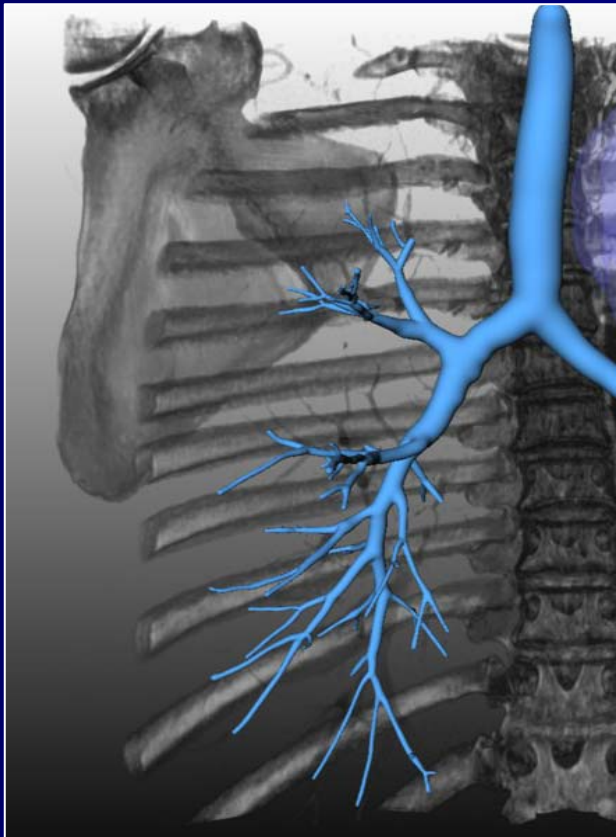
Ground-Glass & Reversibility...Look at the Airways!



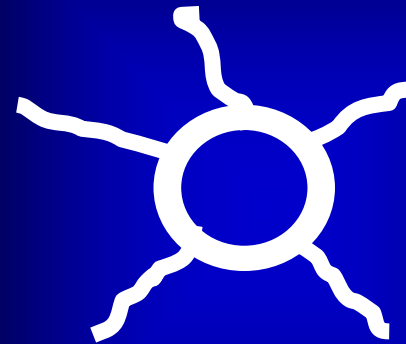
Ground-Glass Opacification

Tractional Dilatation of Airways

Normal Tapering



Alveolar Attachments



FROM: Google Images

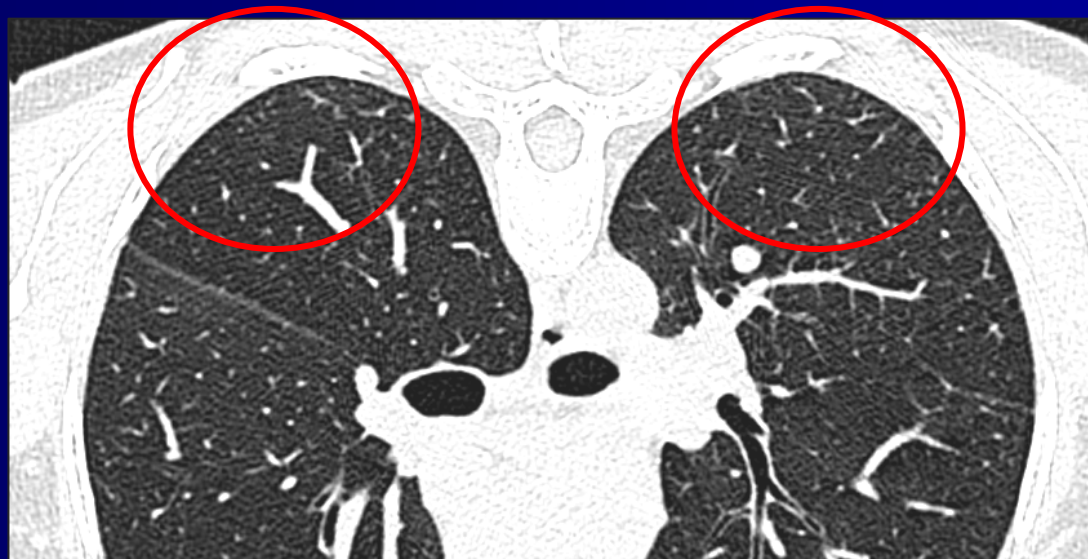
SURVIVAL GUIDE

Rule 4:

**TRY NOT TO
"PLEASE" THE CLINICIAN!**

Importance of Clinical Data

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 COMMOM / IMPORTANT
DILDs**

HRCT diagnoses

Degree of Diagnostic Confidence /Accuracy

Idiopathic pulmonary fibrosis / CFA

++++/-

Langerhans' cell histiocytosis

+++/-

Asbestosis

+++/-

Sarcoidosis

+++/-

Extrinsic allergic alveolitis

++/-

Alveolar proteinosis

++/-

(Cryptogenic) organizing pneumonia

+/--

Non-specific interstitial pneumonia

+/--

Smoking-related ILD

+/--

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

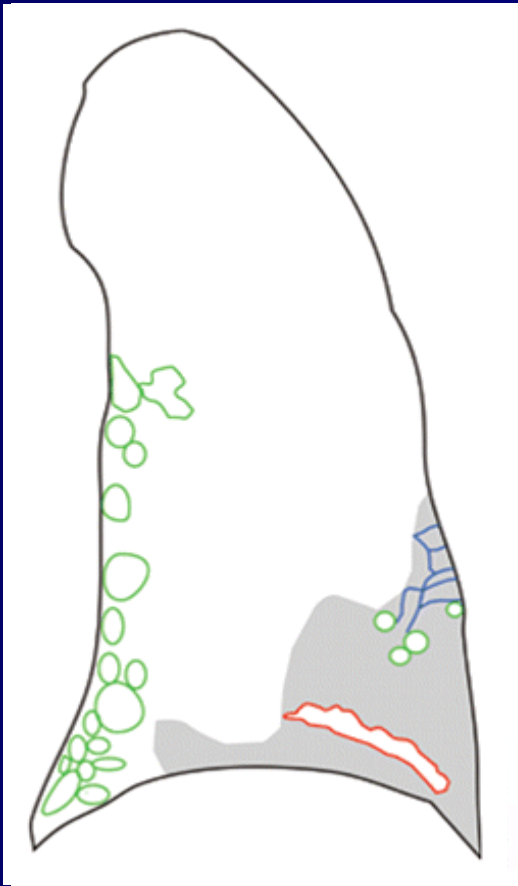
NB Radiologist / pathologist makes diagnosis of pattern ... not synonymous with CFA / IPF

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

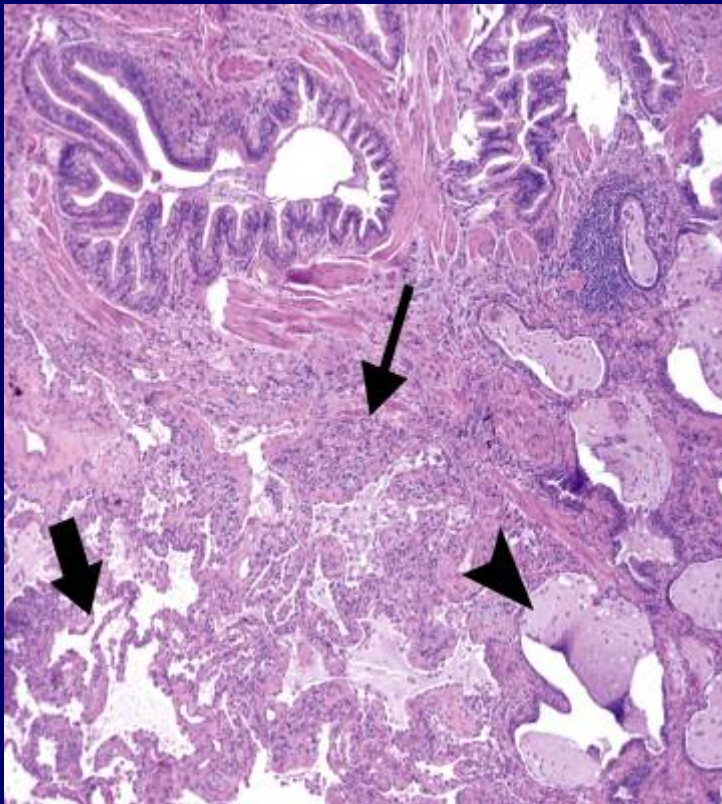
Temporal and Spatial Heterogeneity

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:

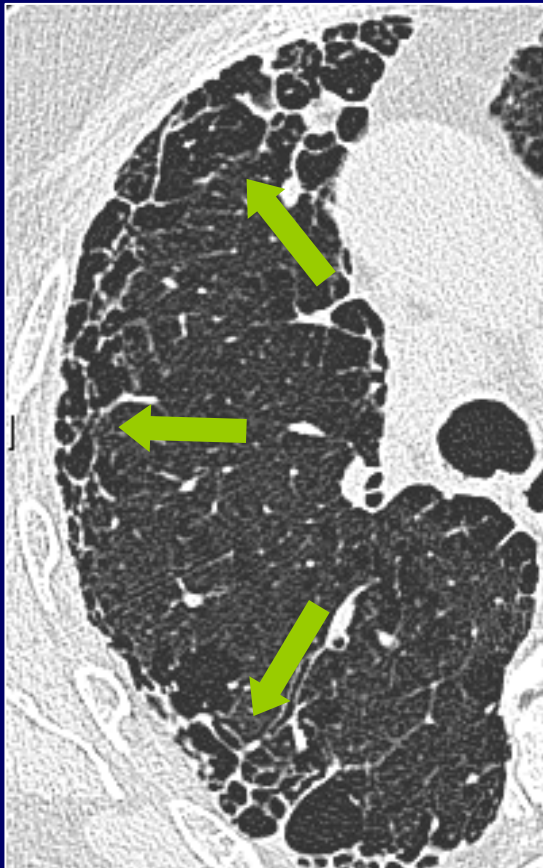
Mueller-Mang et al. What every radiologist should know about idiopathic interstitial pneumonias *Radiographics* 2007;27:595-615

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

Idiopathic Pulmonary Fibrosis

Characteristic HRCT Features

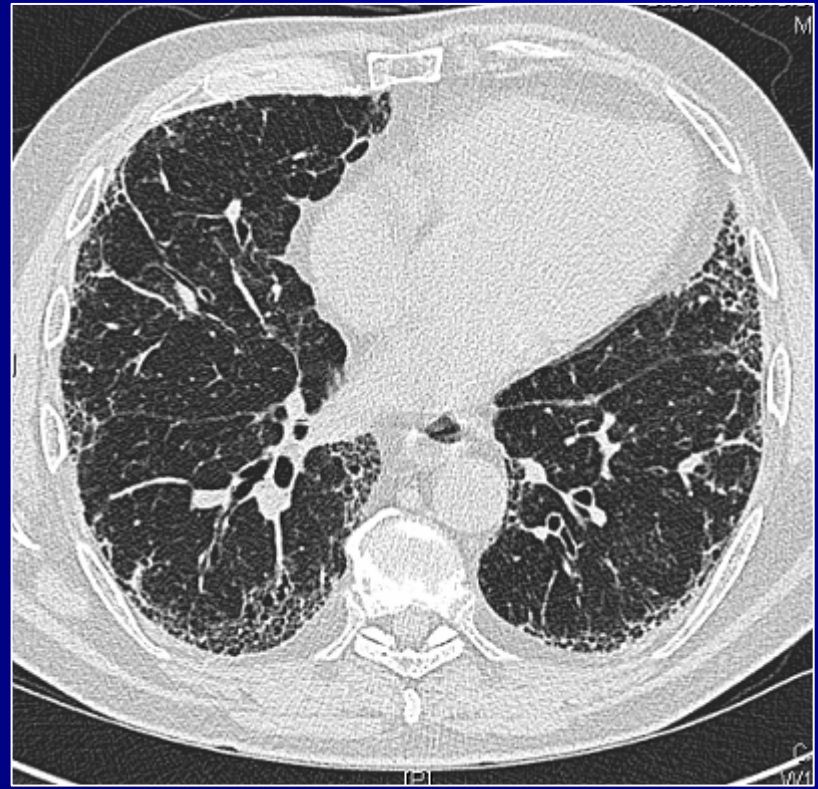
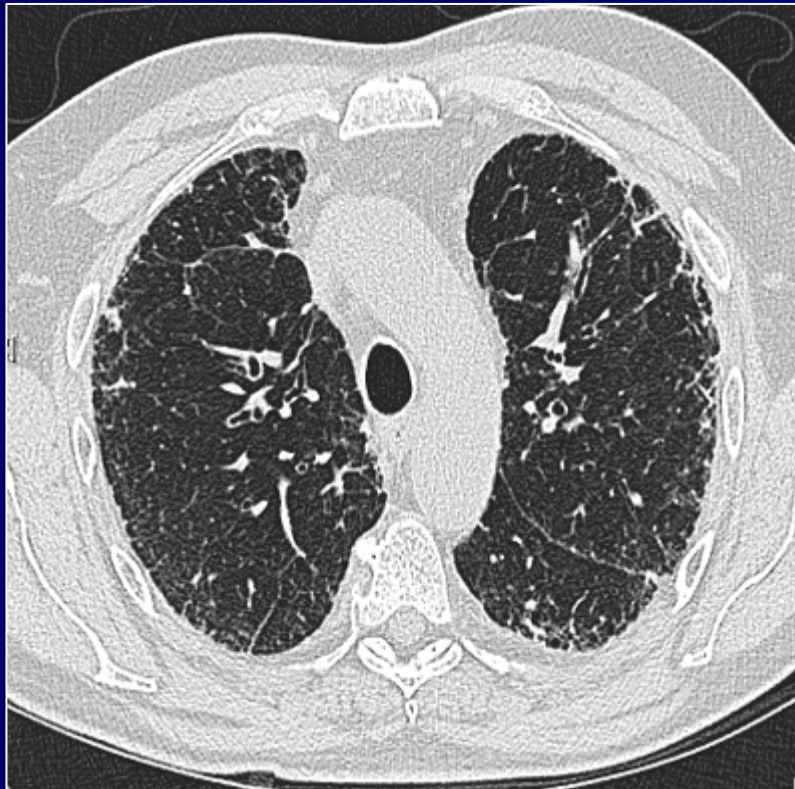
RETICULAR PATTERN WITH HONEYCOMBING



Idiopathic Pulmonary Fibrosis

Characteristic HRCT Features

UPPER ZONES vs LOWER ZONES



Idiopathic Pulmonary Fibrosis

Characteristic HRCT Features

- Reticular pattern *with* honeycombing
- Basal
- Subpleural

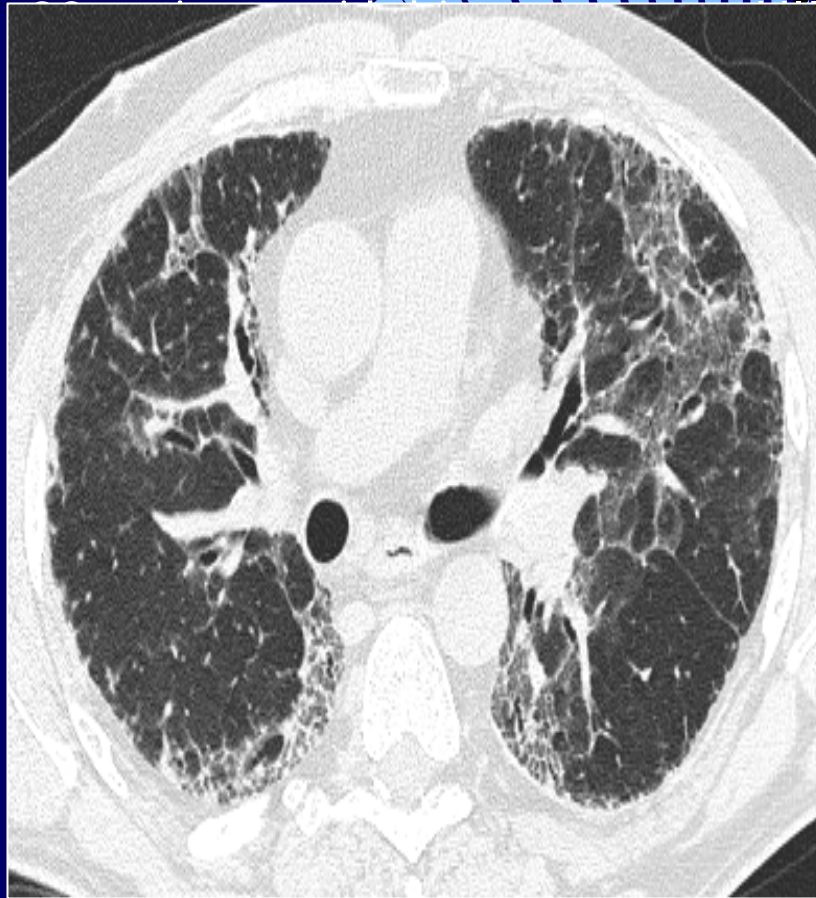
High (>95%) positive predictive value of an (experienced) confident CT diagnosis^{1,2}

¹ Hunninghake et al. Utility of a Lung Biopsy for the Diagnosis of Idiopathic Pulmonary Fibrosis. *Am J Respir Crit Care Med* 2001;164:193

² Flaherty et al. Radiological versus Histological Diagnosis of UIP and NSIP: Survival Implications. *Thorax* 2003;58:143

Idiopathic Pulmonary Fibrosis

Atypical HRCT Features



Langerhans' Cell Histiocytosis

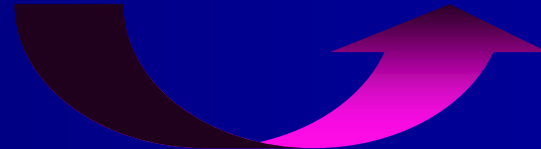
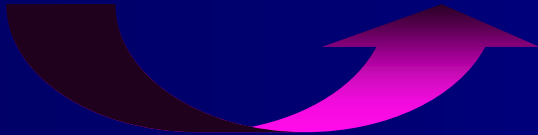
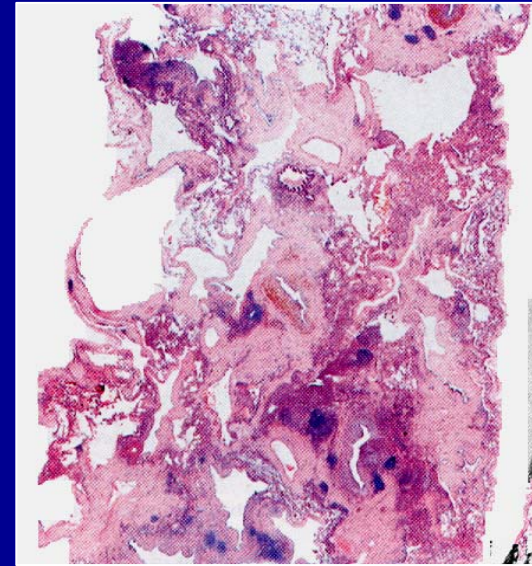
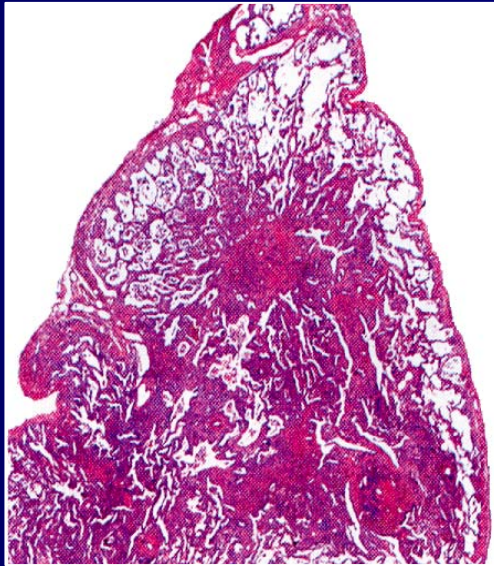
Langerhans' Cell Histiocytosis

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

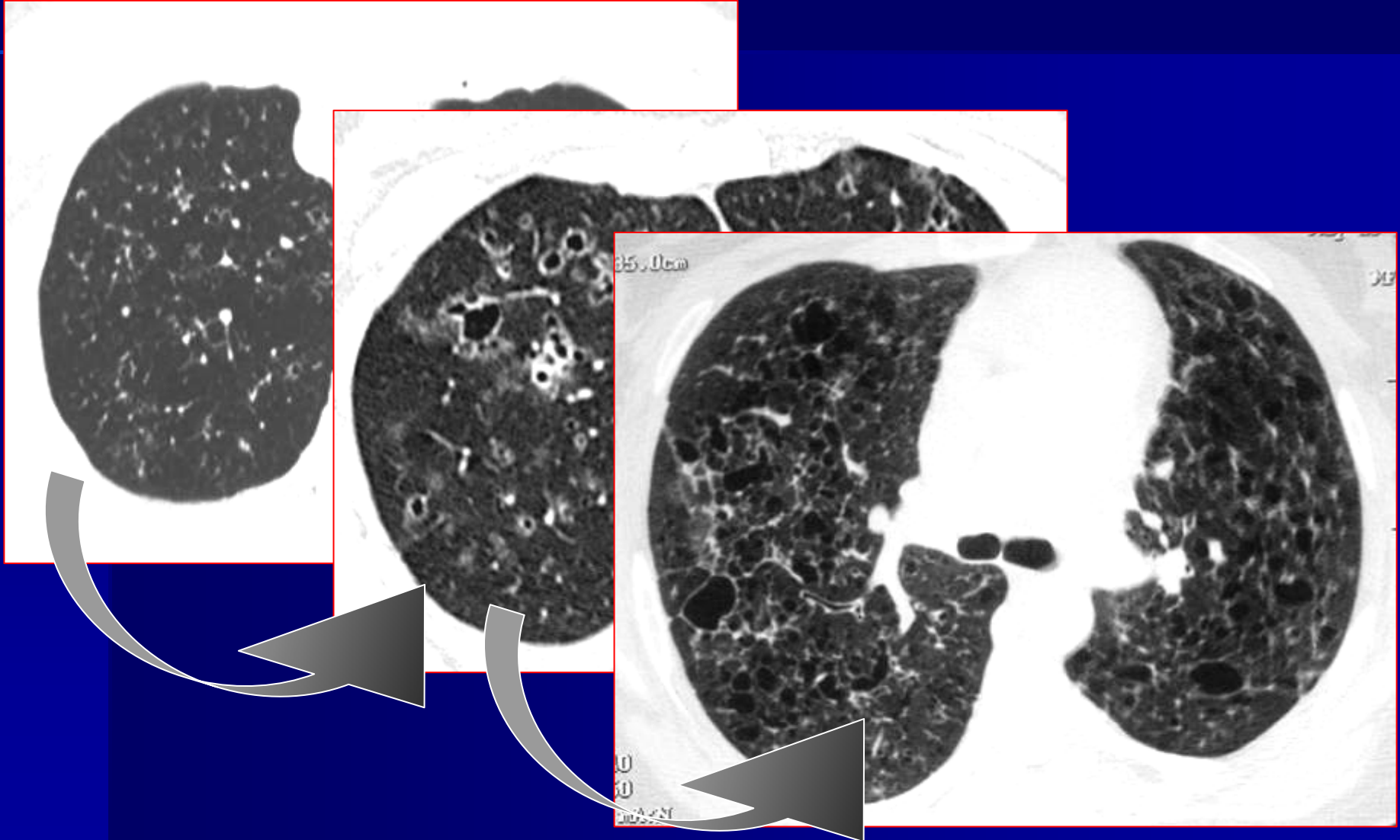
Langerhans' Cell Histiocytosis

Histopathological Changes

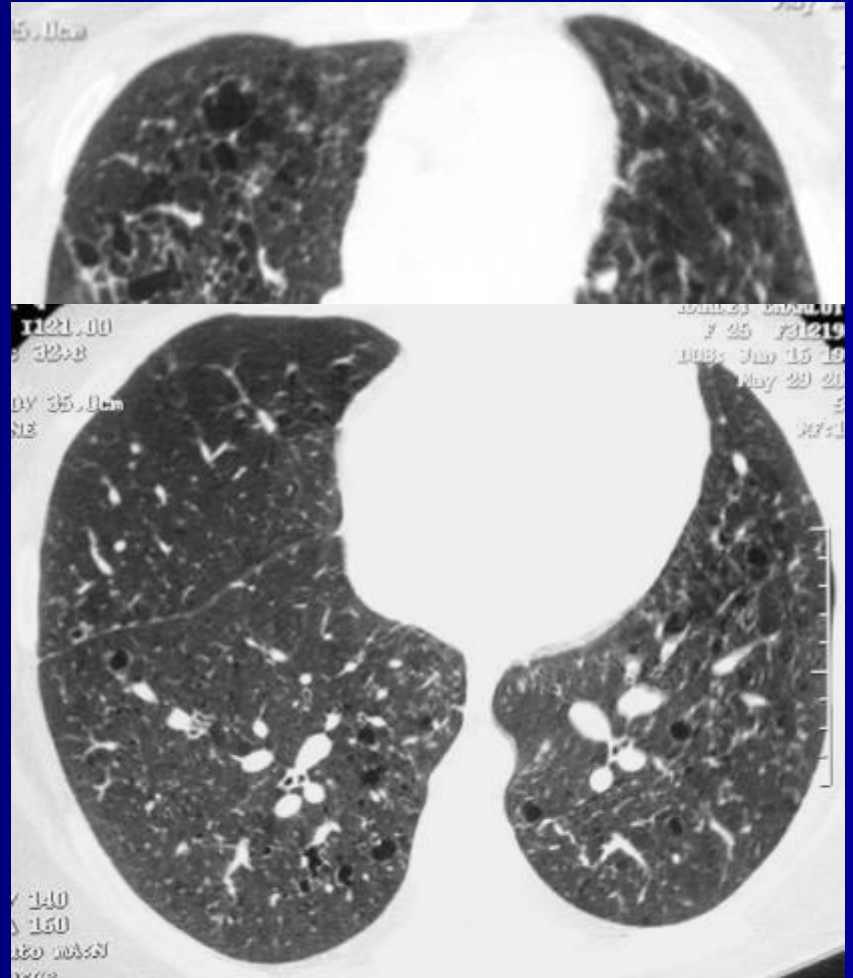
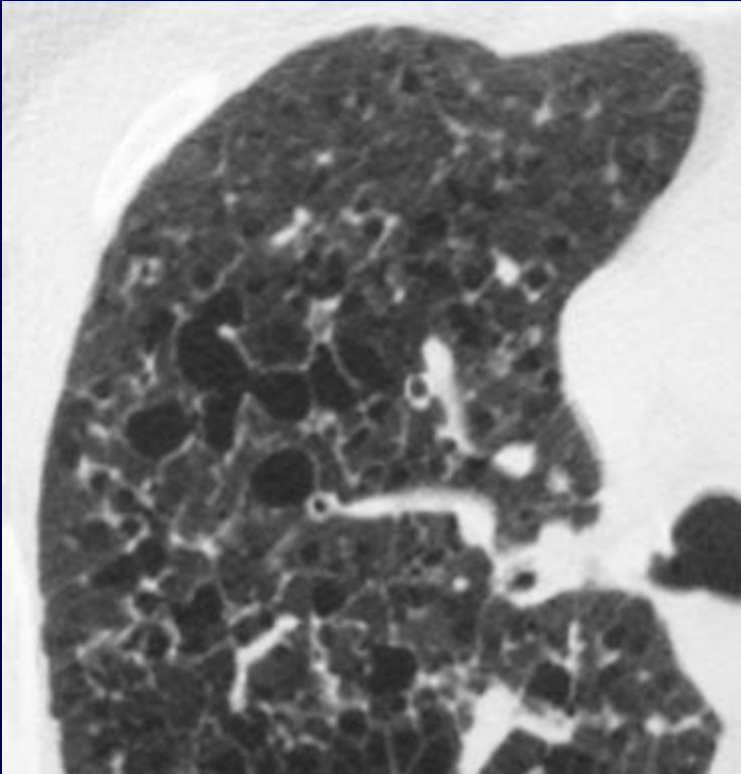


Langerhans' Cell Histiocytosis

Serial HRCT Changes



"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 pneumatocoeles

Sarcoidosis

Sarcoidosis

Essential Pathology

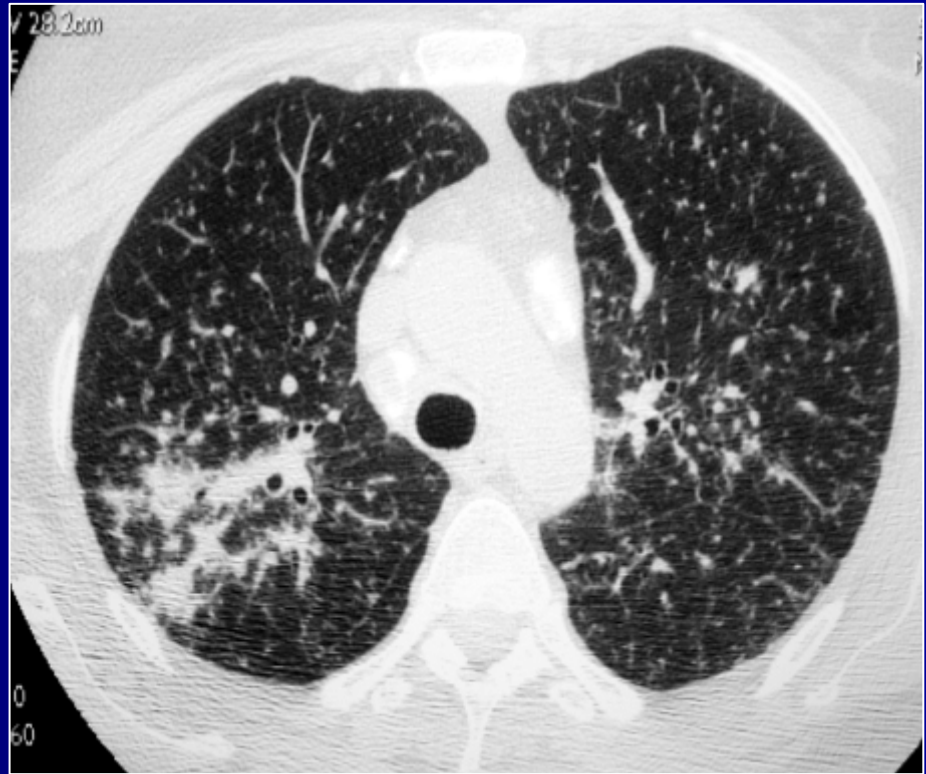
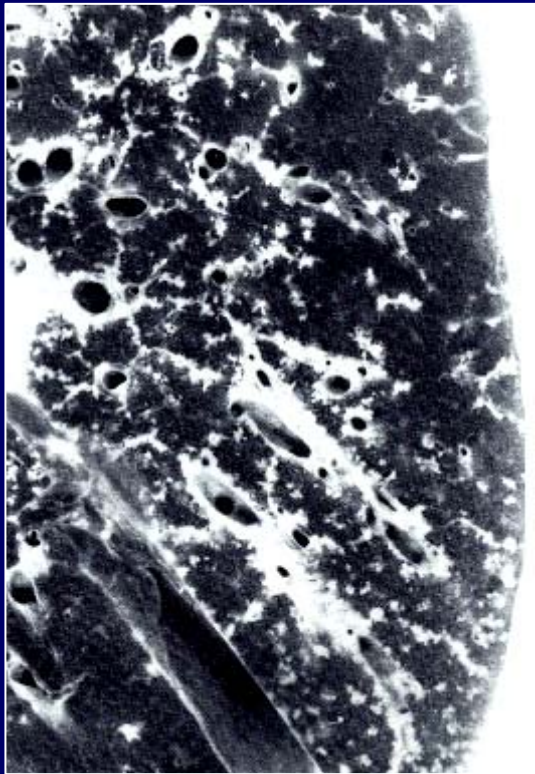
- Mononuclear alveolitis
- Early, loosely formed granulomas
- Along lymphatic pathways
- Tightly packed N-C granulomas
epithelioid histiocytes, multinucleate giant cells, surrounded by lymphocytes, monocytes and fibroblasts



IMAGES FROM: Corrin B. Restrictive ventilatory impairment. In: *Pathology of the Lungs*. Corrin B (ed). Churchill Livingstone, London (2000)

Sarcoidosis

Pathological vs Radiological Features

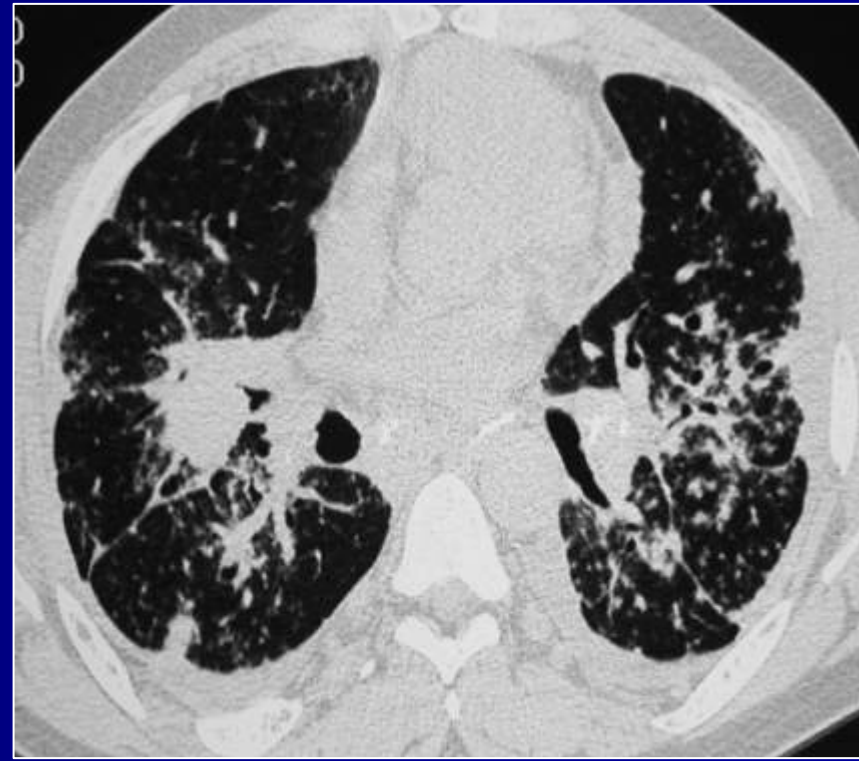
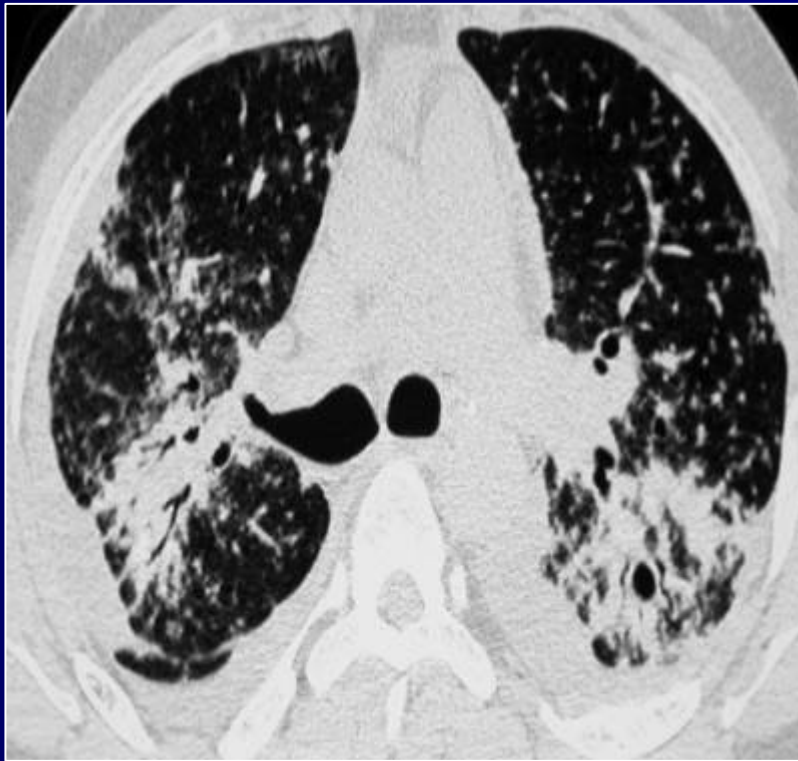


PATHOLOGY IMAGE FROM: Thurlbeck WM & Churg AM. Interstitial Lung Diseases. In: *Pathology of the Lung*. (2nd edition). Thurlbeck WM & Churg AM (eds). Thieme, New York (1995)

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

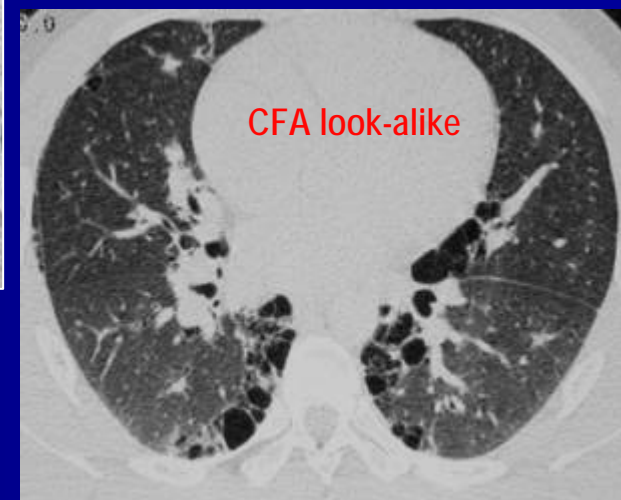
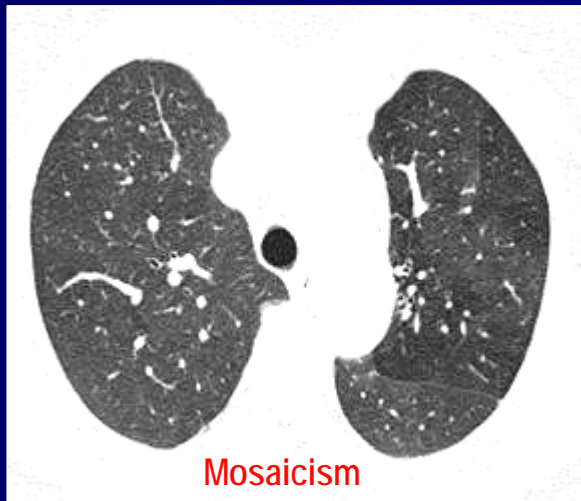
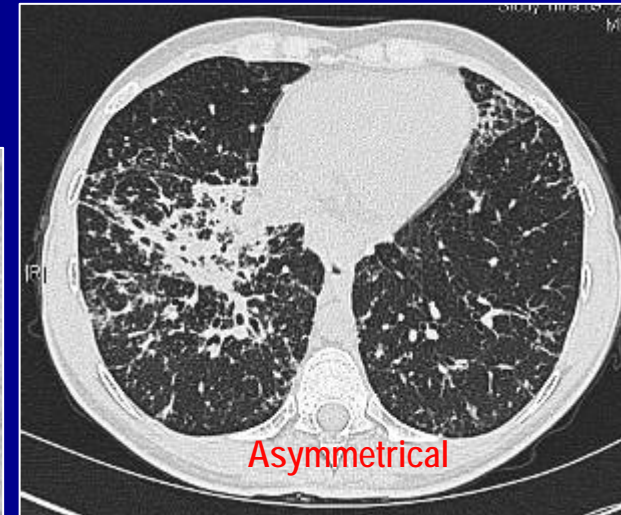
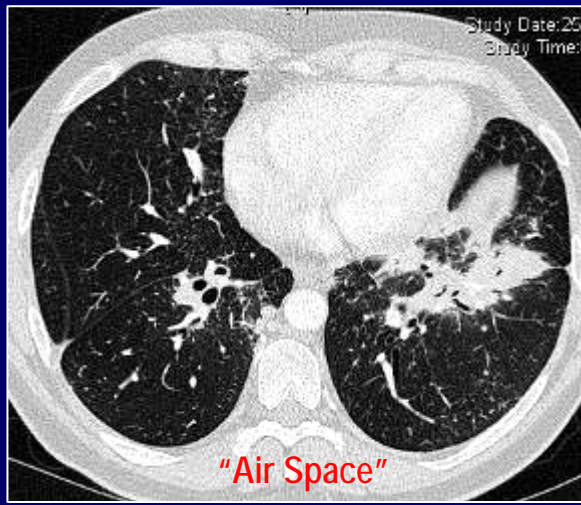
Sarcoidosis

HRCT Features



Sarcoidosis

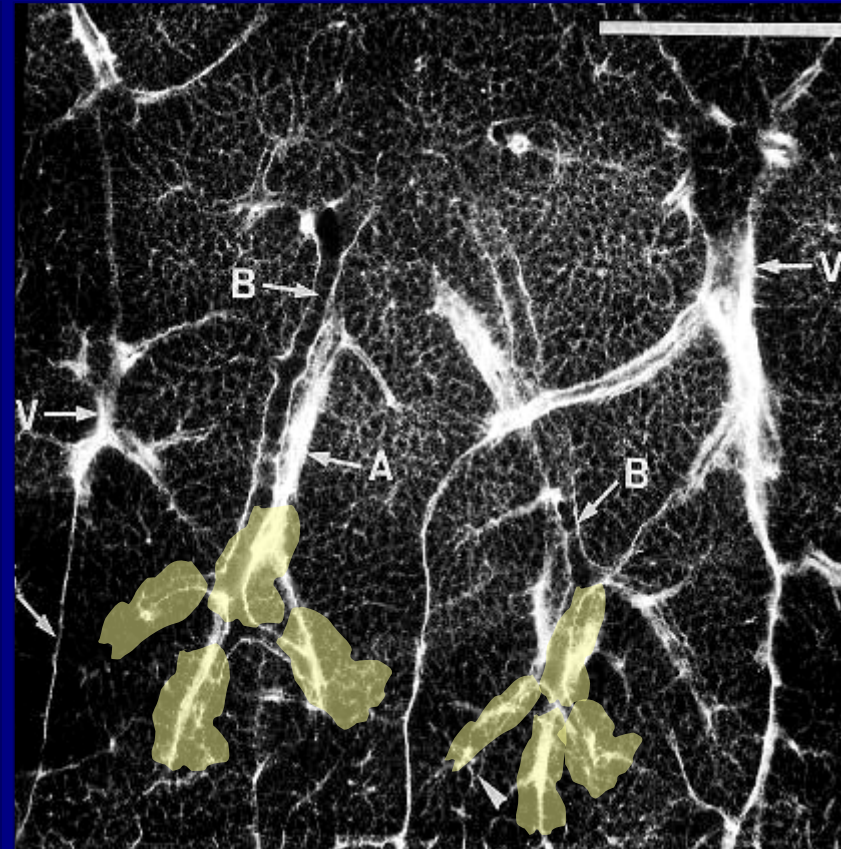
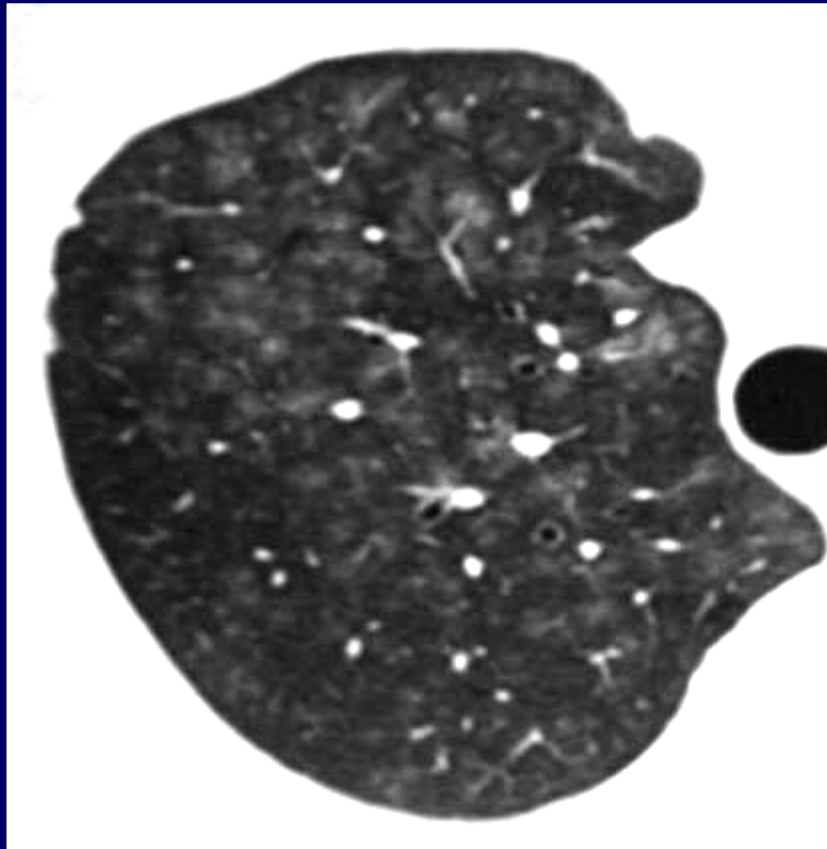
Unusual Patterns



Extrinsic Allergic Alveolitis

Extrinsic Allergic Alevolitis

Radiological-Pathological Relationships



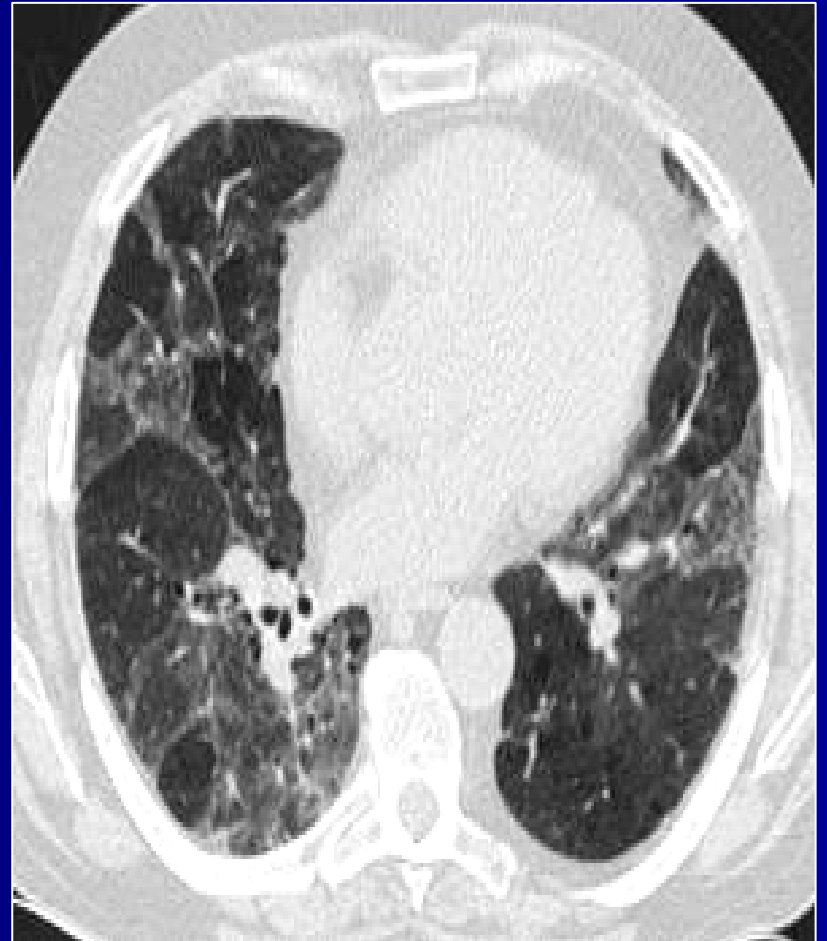
Extrinsic Allergic Alevolitis

Subacute Disease



Extrinsic Allergic Alevolitis

Chronic Disease ... "Head Cheese" Sign



Alveolar Proteinosis

Alveolar Proteinosis

The "Crazy Paving" Sign

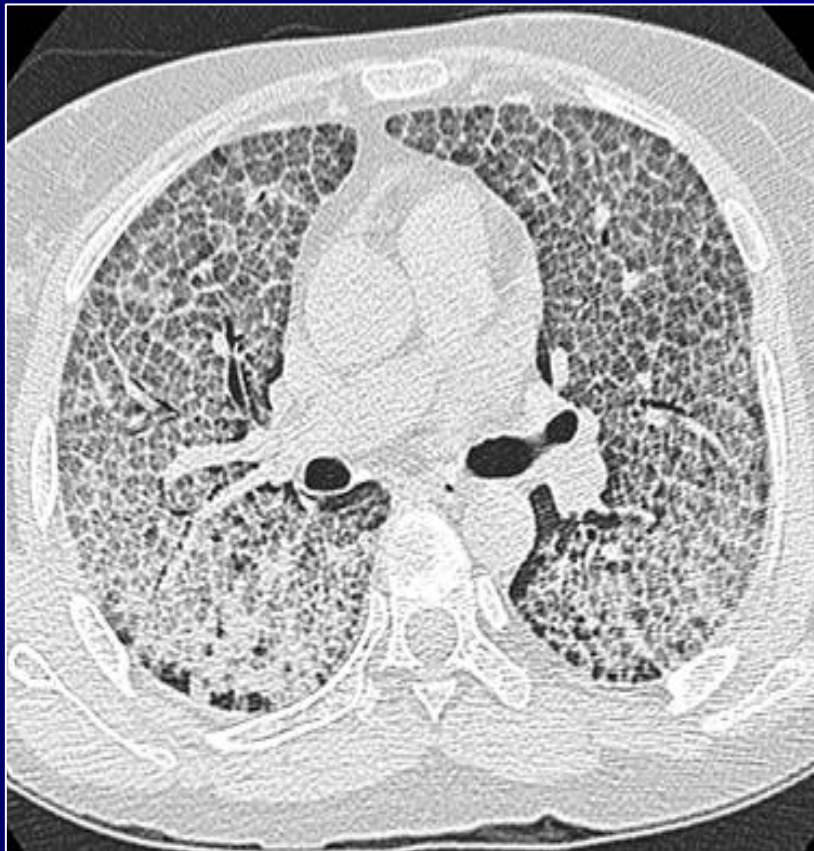


IMAGE COURTESY: Dr Susan J Copley,
Hammersmith Hospital, London

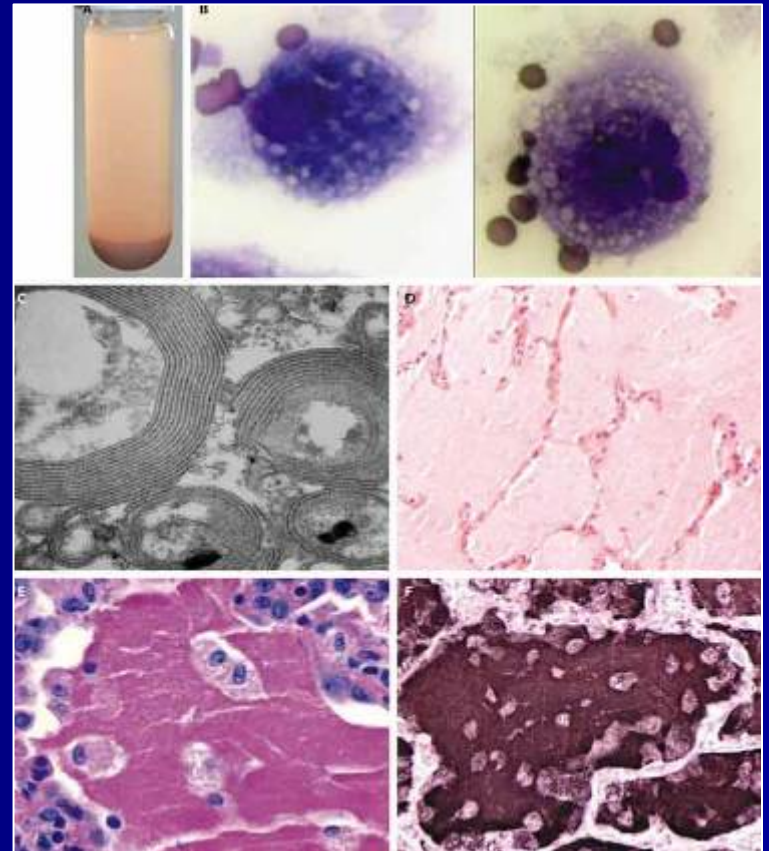
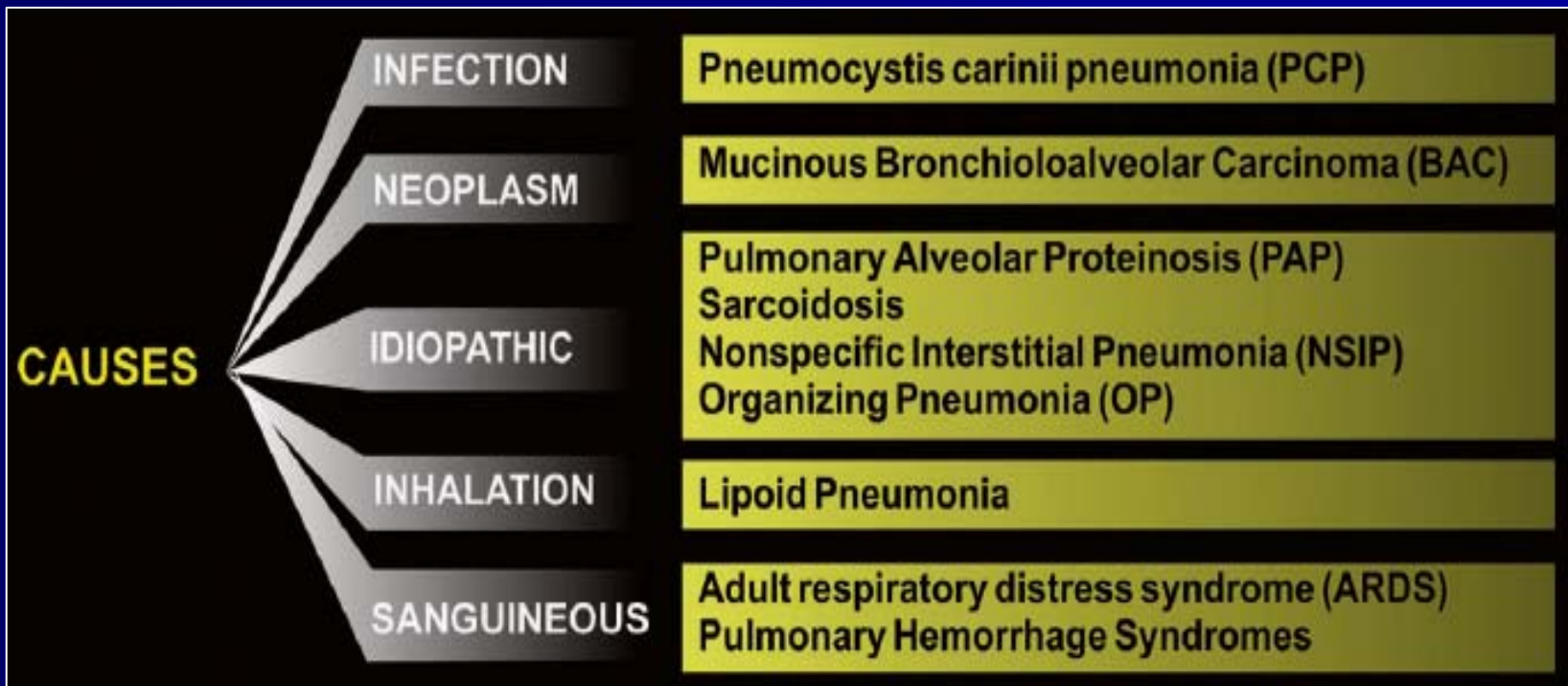


IMAGE FROM: Trapnell BC et al. Pulmonary alveolar proteinosis. *N Engl J Med* 2003; 349-2527

Alveolar Proteinosis

The "Crazy Paving" Sign



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

ESTI2012

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20TH ANNUAL MEETING
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