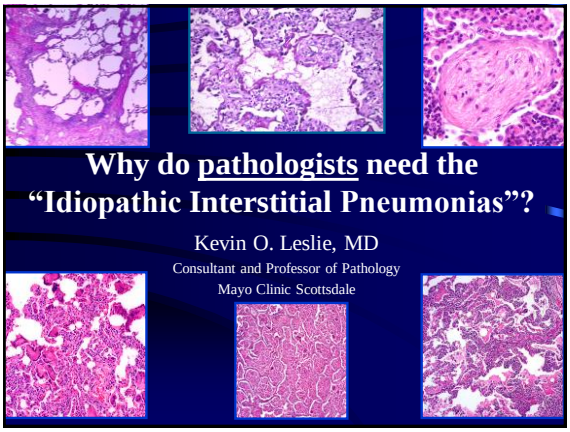


Why do pathologists need the "Idiopathic Interstitial Pneumonias"?

Kevin O. Leslie, MD
Consultant and Professor of Pathology
Mayo Clinic Scottsdale

Why do we classify diseases?

- To define expected natural history
- To select optimal therapy
- To try to understand etiology
- To try to understand pathogenesis

The current state of the art for treatment of "ILD"

1. Antibiotics
2. Bronchodilators and anti-tussives
3. Immunosuppression
4. Small molecule therapy for IPF (perfenidone and ninteninab)
5. PHT disease modifying agents
6. Transplantation

What makes a disease idiopathic?

- Absence of an identifiable cause…

…..after exhaustive searching!

What makes a disease idiopathic?

Therefore…

PATHOLOGISTS CANNOT DIAGNOSE

IDIOPATHIC DISEASE!

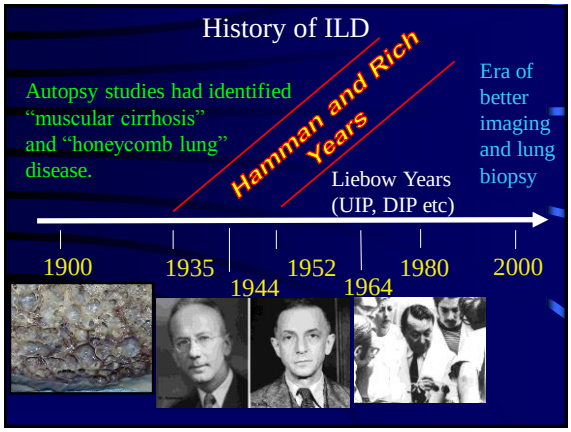
The World Of ILD

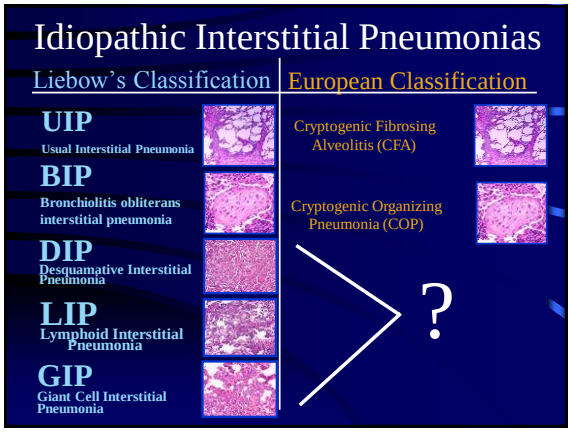
Diffuse lung diseases of "known" etiology

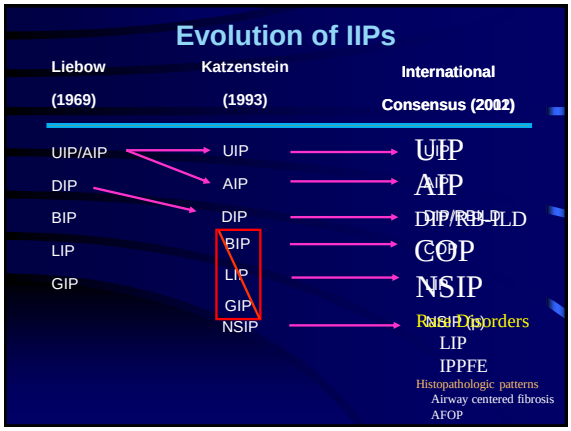
Sarcoidosis/Berylliosis	Diseases related to CVD	Organizing pneumonia
Hypersensitivity	Diseases related to drugs	Amyloidosis
Eosinophilic pneumonia	Pneumoconioses	Lymphangitic tumor
RBILD & Smokers ILD	Alveolar proteinosis	Vasculitis/DAH
Diffuse alveolar damage	Constrictive bronchiolitis	

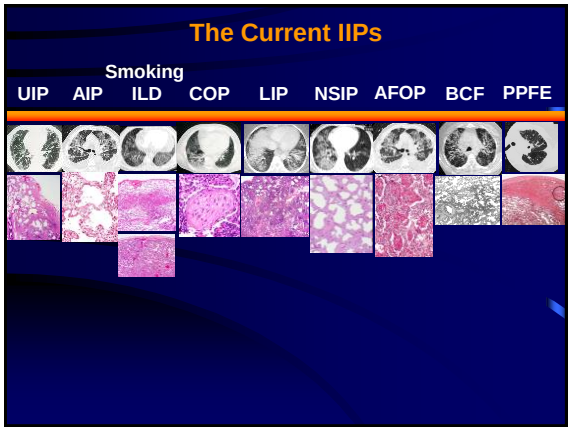
Diffuse lung diseases of "unknown" etiology (idiopathic)

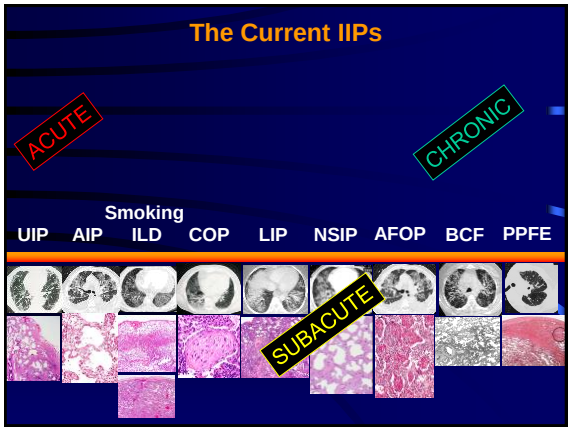
Usual interstitial pneumonia (UIP)	Idiopathic pleuroparenchymal fibroelastosis (IPPFE)
Desquamative interstitial pneumonia (DIP)	Lymphocytic interstitial pneumonia (LIP)
Respiratory bronchiolitis ILD (RB-UILD)	Histopathologic patterns (BCF, AFOP)
Non-specific interstitial pneumonia (NSIP)	
Cryptogenic organizing pneumonia (COP)	
Acute interstitial pneumonia (AIP)	

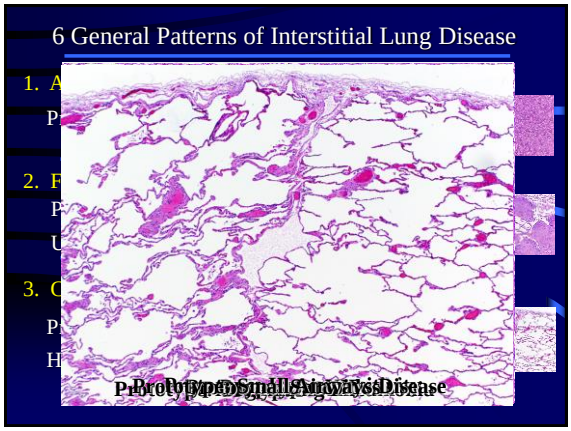

History of ILD
Autopsy studies had identified "muscular cirrhosis" and "honeycomb lung" disease.
Hamman and Rich Years
Era of better imaging and lung biopsy
Liebow Years (UIP, DIP etc)
1900
1935
1944
1952
1964
1980
2000


Idiopathic Interstitial Pneumonias
Liebow's Classification
European Classification
UIP
Usual Interstitial Pneumonia
BIP
Bronchiolitis obliterans interstitial pneumonia
DIP
Desquamative Interstitial Pneumonia
LIP
Lymphoid Interstitial Pneumonia
GIP
Giant Cell Interstitial Pneumonia
Cryptogenic Fibrosing Alveolitis (CFA)
Cryptogenic Organizing Pneumonia (COP)
?


Evolution of IIPs
Liebow (1969)
Katzenstein (1993)
International Consensus (2002)
UIP/AIP
DIP
BIP
LIP
GIP
UIP
AIP
DIP
BIP
LIP
GIP
NSIP
UIP
AIP
DIP/RB-ILD
COP
NSIP
LIP
IPPFE
Histopathologic patterns
Airway centered fibrosis
AFOP


The Current IIPs
Smoking
UIP
AIP
ILD
COP
LIP
NSIP
AFOP
BCF
PPFE


The Current IIPs
ACUTE
CHRONIC
Smoking
UIP
AIP
ILD
COP
LIP
NSIP
AFOP
BCF
PPFE
SUBACUTE


6 General Patterns of Interstitial Lung Disease
1.
2.
3.

6 Patterns of Interstitial Lung Disease

1. Acute Lung Injury
- with hyaline membranes?
- with eosinophils?
- with necrosis?
- with siderophages?

2. Fibrosis
- with temporal heterogeneity?
- with honeycombing?
- with diffuse fibrosis?
- with pleuritis?

3. Cellular Infiltrates
- with lymphocytes and plasma cells?
- with neutrophils?
- with granulomas?
- with focal organization?
- with pleuritis?

4. Alveolar Filling
- with macrophages?
- with neutrophils?
- with organizing pneumonia?
- with eosinophilic material?
- with hemorrhage?

5. Nodules
- with granulomas?
- with lymphoid tissue?
- with necrosis?
- with atypical cells?
- with stellate scars?
- with organizing pneumonia?

6. Minimal Changes
- with small airways disease?
- with vascular changes?
- with cysts?
- sampling problem?

6 Patterns of Interstitial Lung Disease

1. Acute Lung Injury
Infections! Drug
CVD DAH
AIP AEP AFOP

2. Fibrosis
Chr HP CVD Drug
UIP NSIP IPPFE BrFib

3. Cellular Infiltrates
HP CVD Drug Tox
NSIP LIP

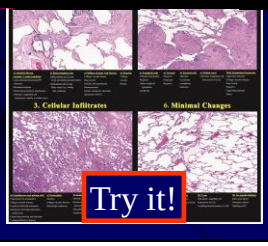
4. Alveolar Filling
Resolving Infections
EP Drugs DAH PAP
RBILD/DIP COP

5. Nodules
Infections Tumors
PLCH WG Chr OP
Sarcoid

6. Minimal Changes
SAD PHT LAM

w/ Hyaline membranes	w/ Eosinophils	w/ Necrosis	w/ Siderophages
Infections	Acute eosinophilic pneumonia	Infections	Diffuse alveolar hemorrhage
Collagen vascular diseases	Drug toxicity	- Viral	Collagen vascular diseases
Drug toxicity	DAD in a smoker	- Bacterial	Drug toxicity
Idiopathic DAD		- Fungal	DAD in a smoker
(acute interstitial pneumonia)		Some tumors	
Clinical ARDS			

Differential diagnosis based on additional findings



Try it!

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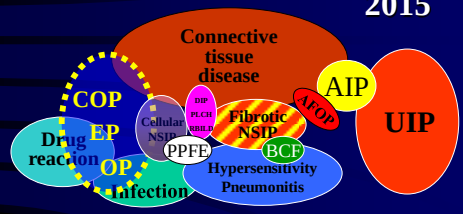
Diagnosis STILL Matters!



Prognostic Classification of ILD