

A Pattern-Based Approach to *Interstitial Lung Disease*

Part 1: Introduction

Kevin O. Leslie, M.D.

Consultant and Professor

Department of Pathology and Laboratory Medicine

Mayo Clinic Arizona

Presenter Disclosures

Kevin O. Leslie, MD

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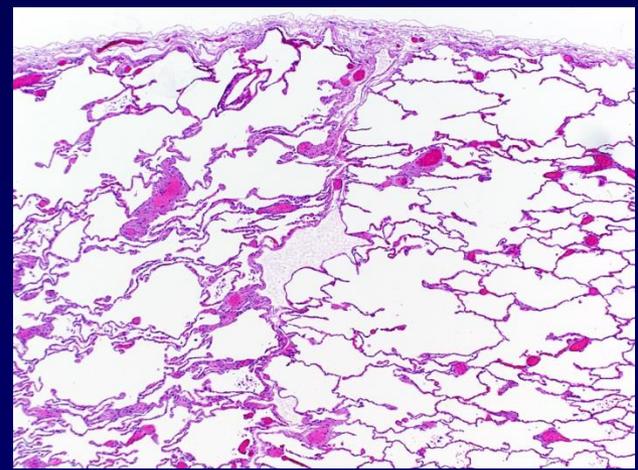
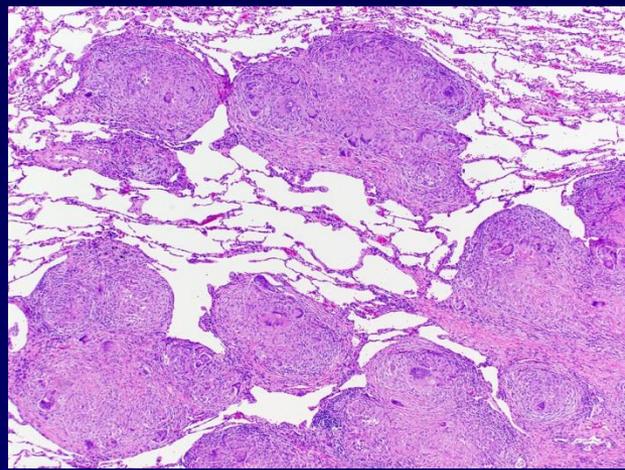
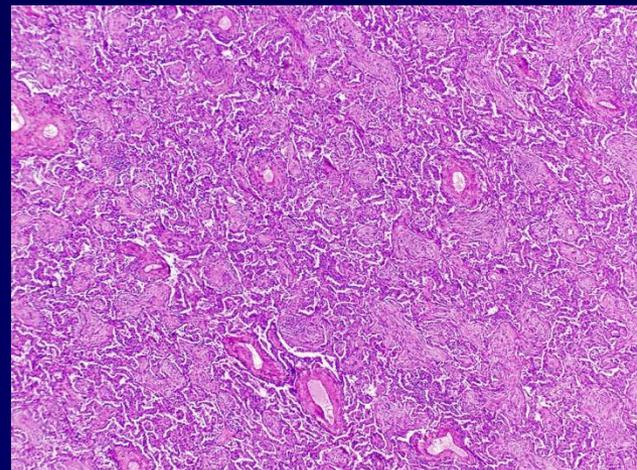
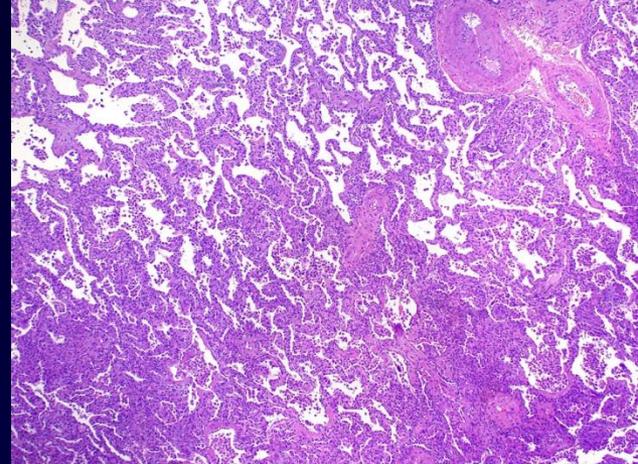
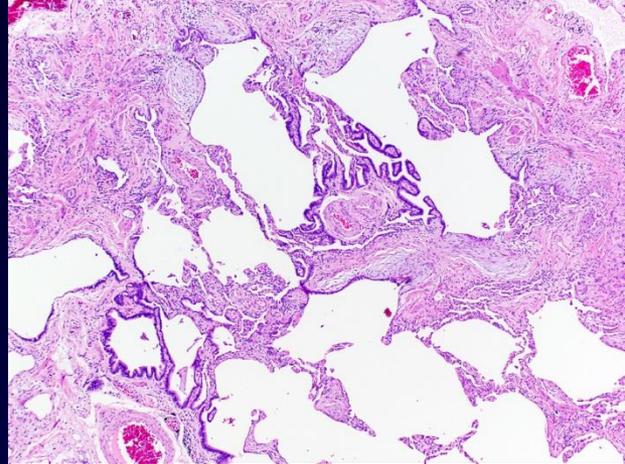
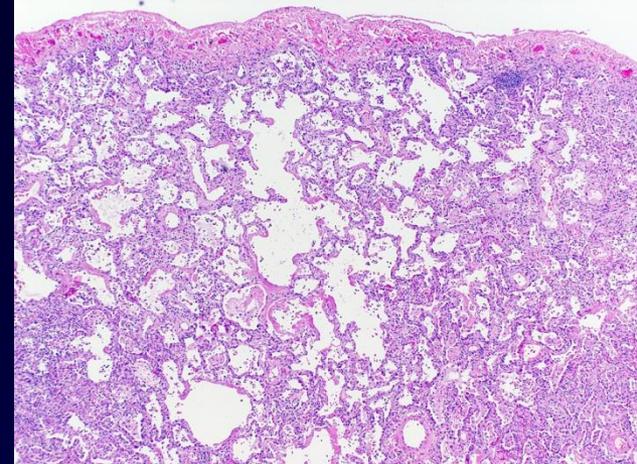
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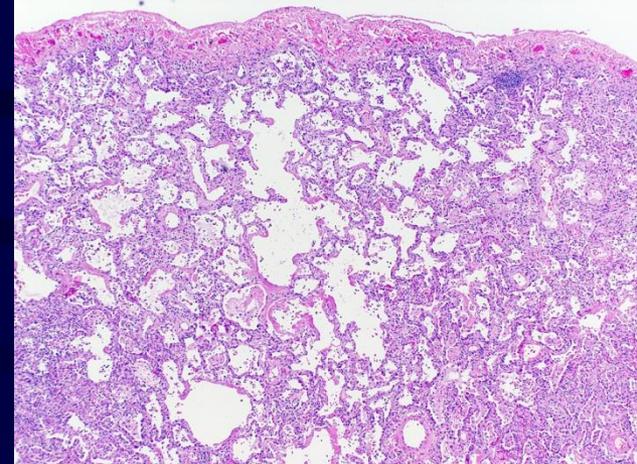
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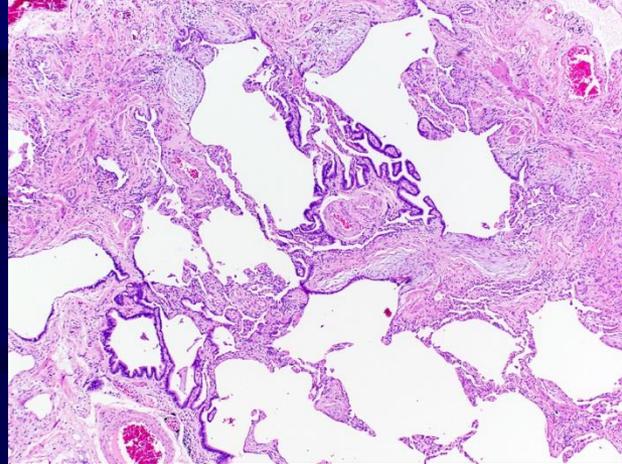
No Disclosures

Pre-Test

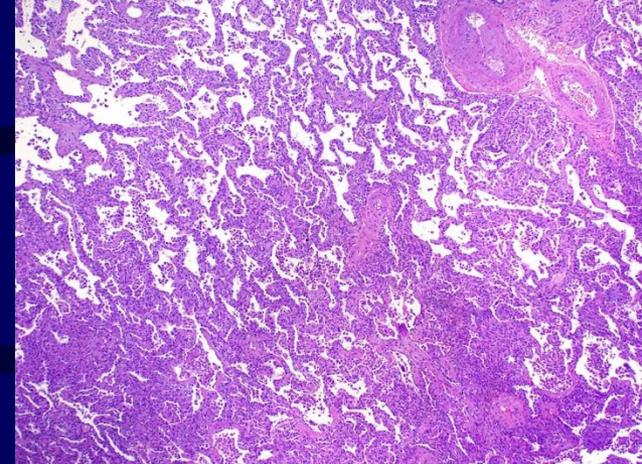




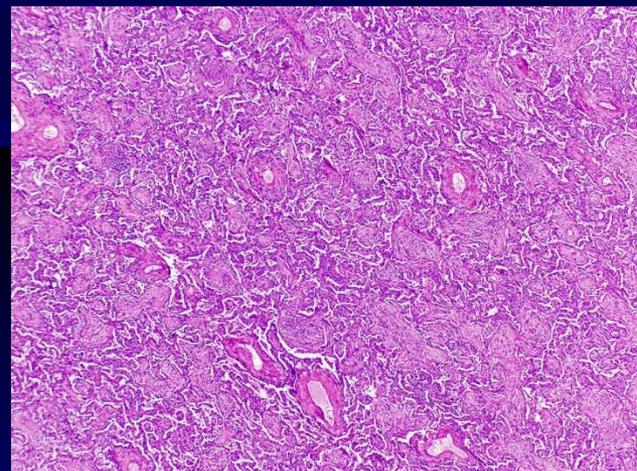
Diffuse Alveolar Damage



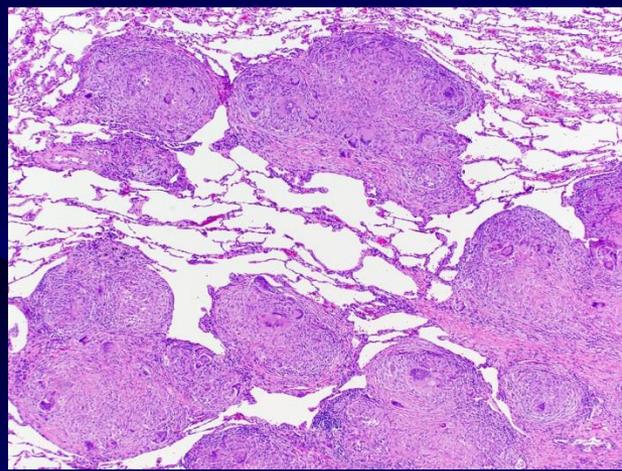
Usual Interstitial Pneumonia



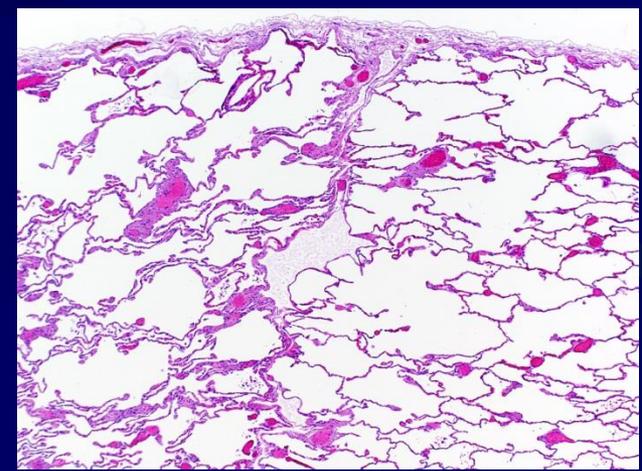
Hypersensitivity Pneum.



Organizing Pneumonia

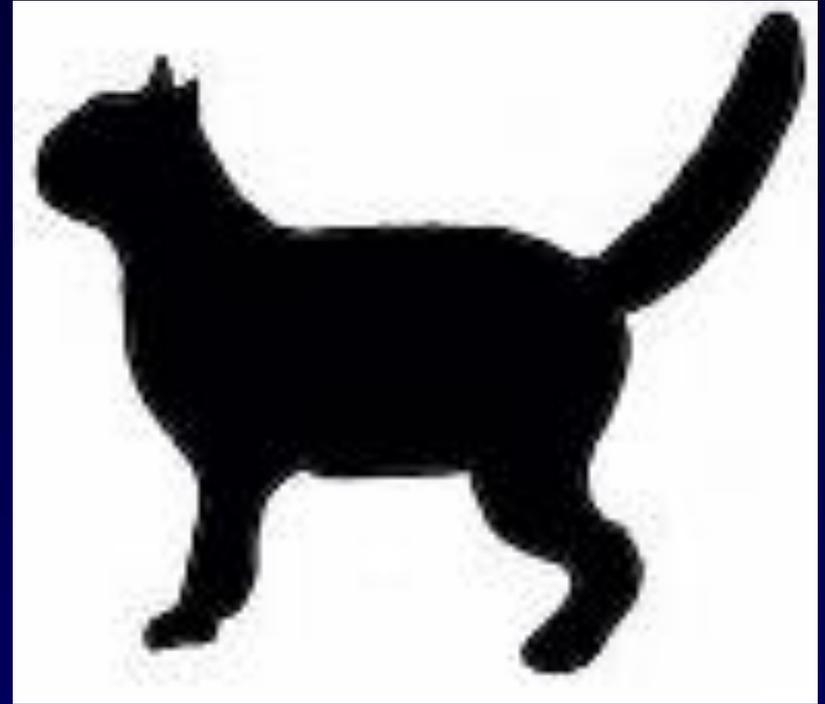


Sarcoidosis



Constrictive Bronchiolitis

What is in a Silhouette?



Objectives

History, Nomenclature, and Classification of ILD

Clinical Patterns of ILD

- Acute, Subacute, Chronic
-

Radiological Patterns of ILD

- Ground glass and consolidation, Fibrosis, Nodules, Cysts and Mosaic Perfusion
-

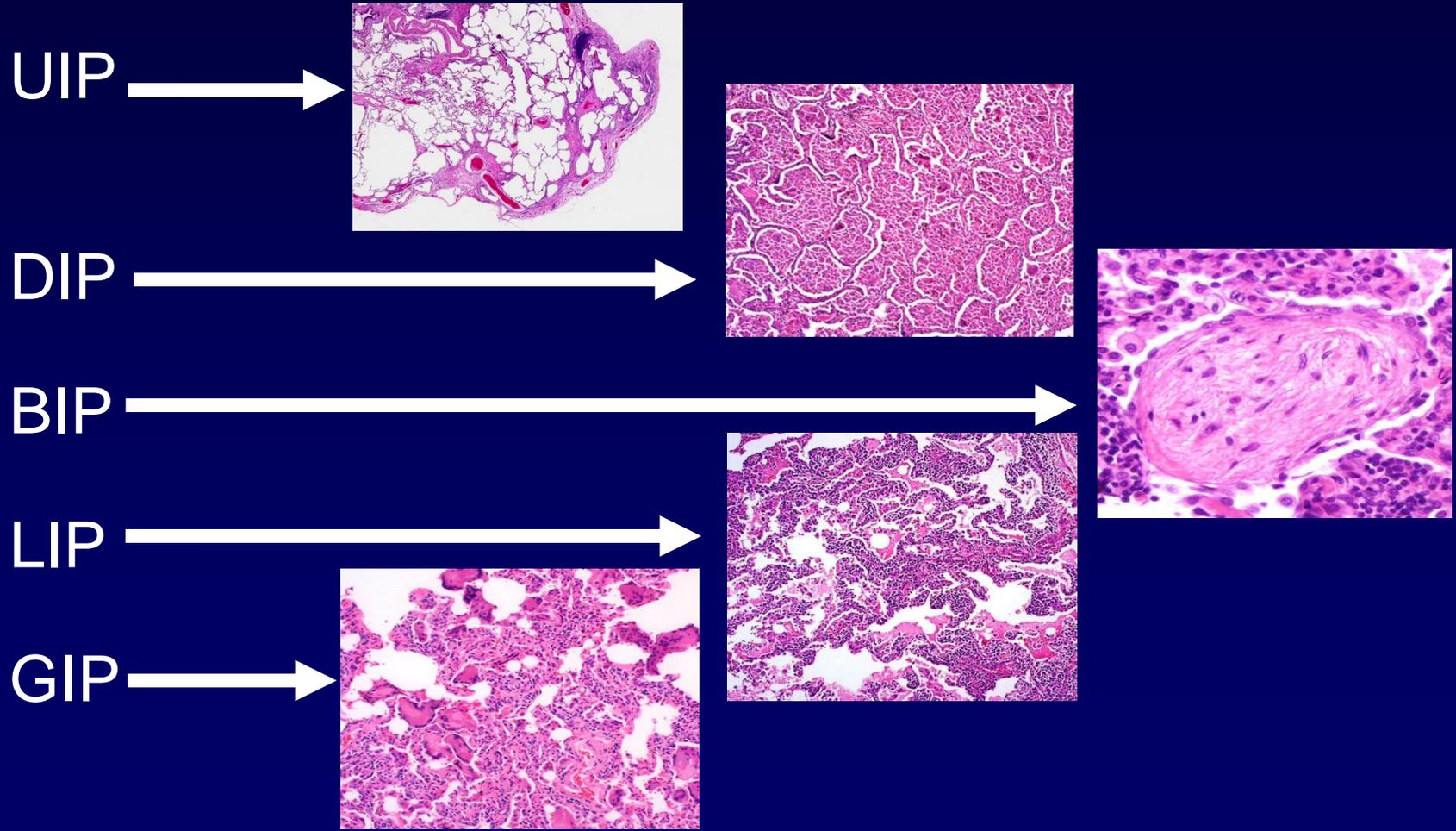
Histopathological Patterns

- Acute injury, Fibrosis, Cellular infiltrates, Alveolar filling, Nodules, Minimal changes

Evolution of Classification

Liebow

(1969)



Evolution of Classification

Liebow

Katzenstein

(1969)

(1993)

UIP



UIP



AIP

DIP



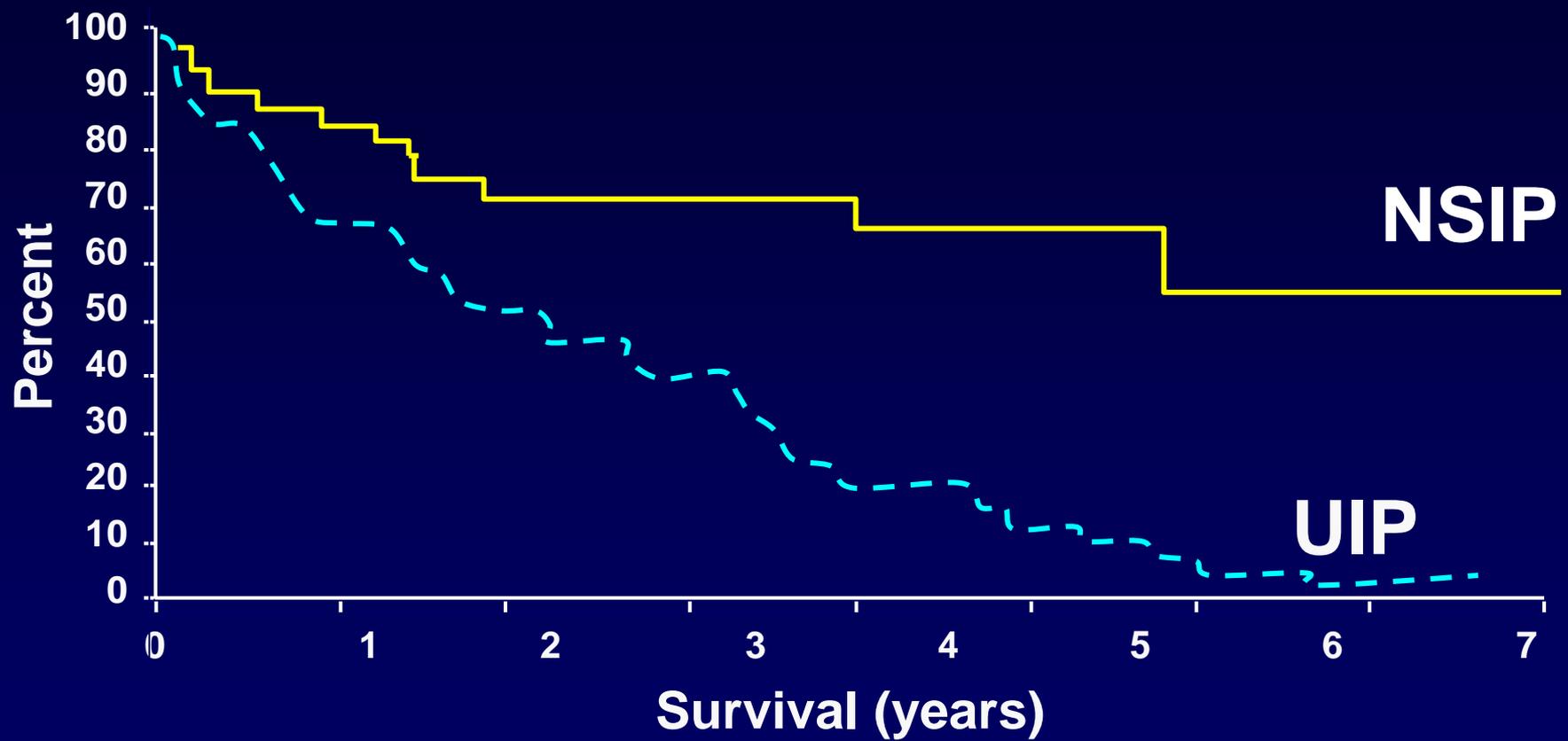
DIP

BOOP

LIP

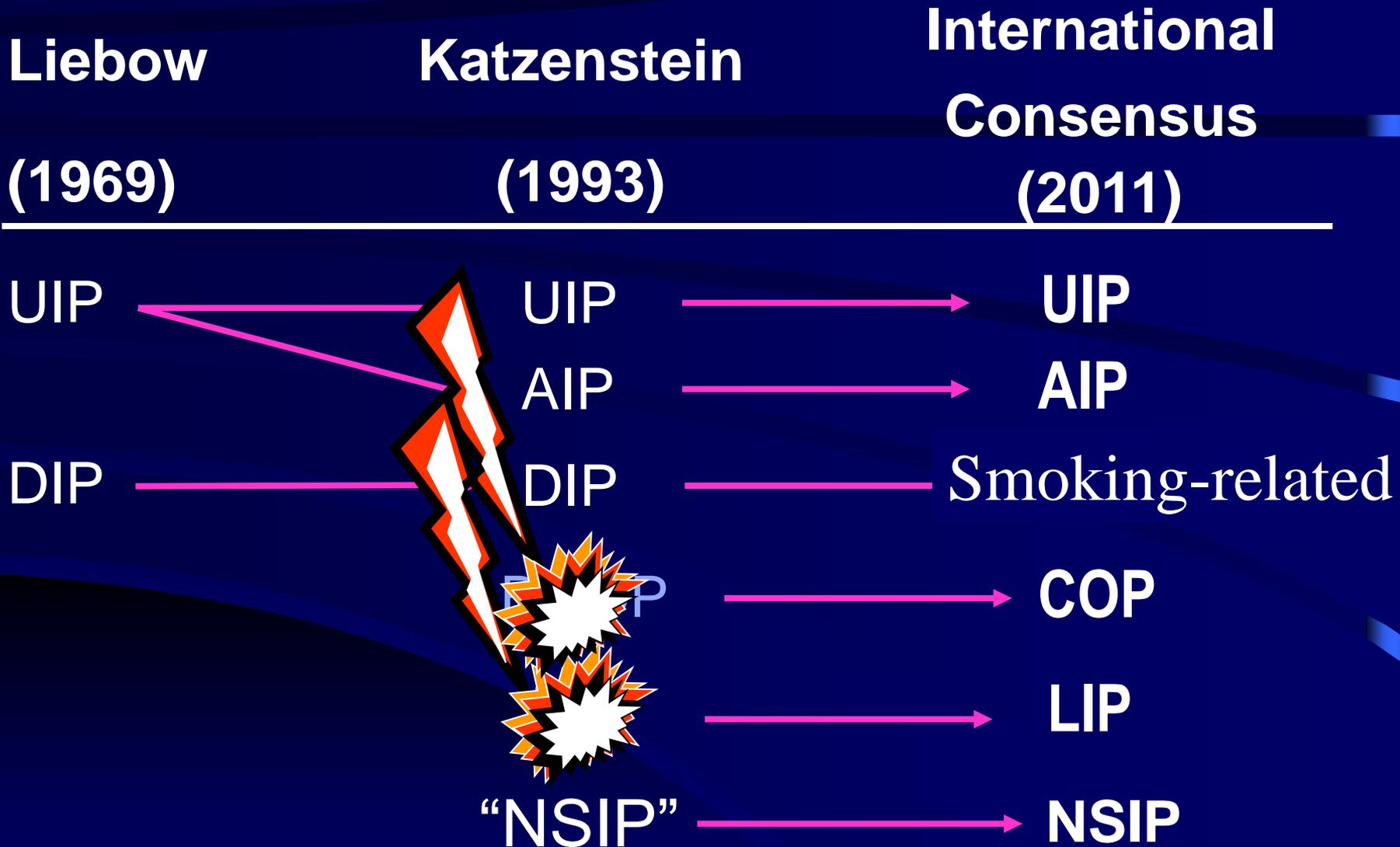
“NSIP”

Subtypes of NSIP vs UIP

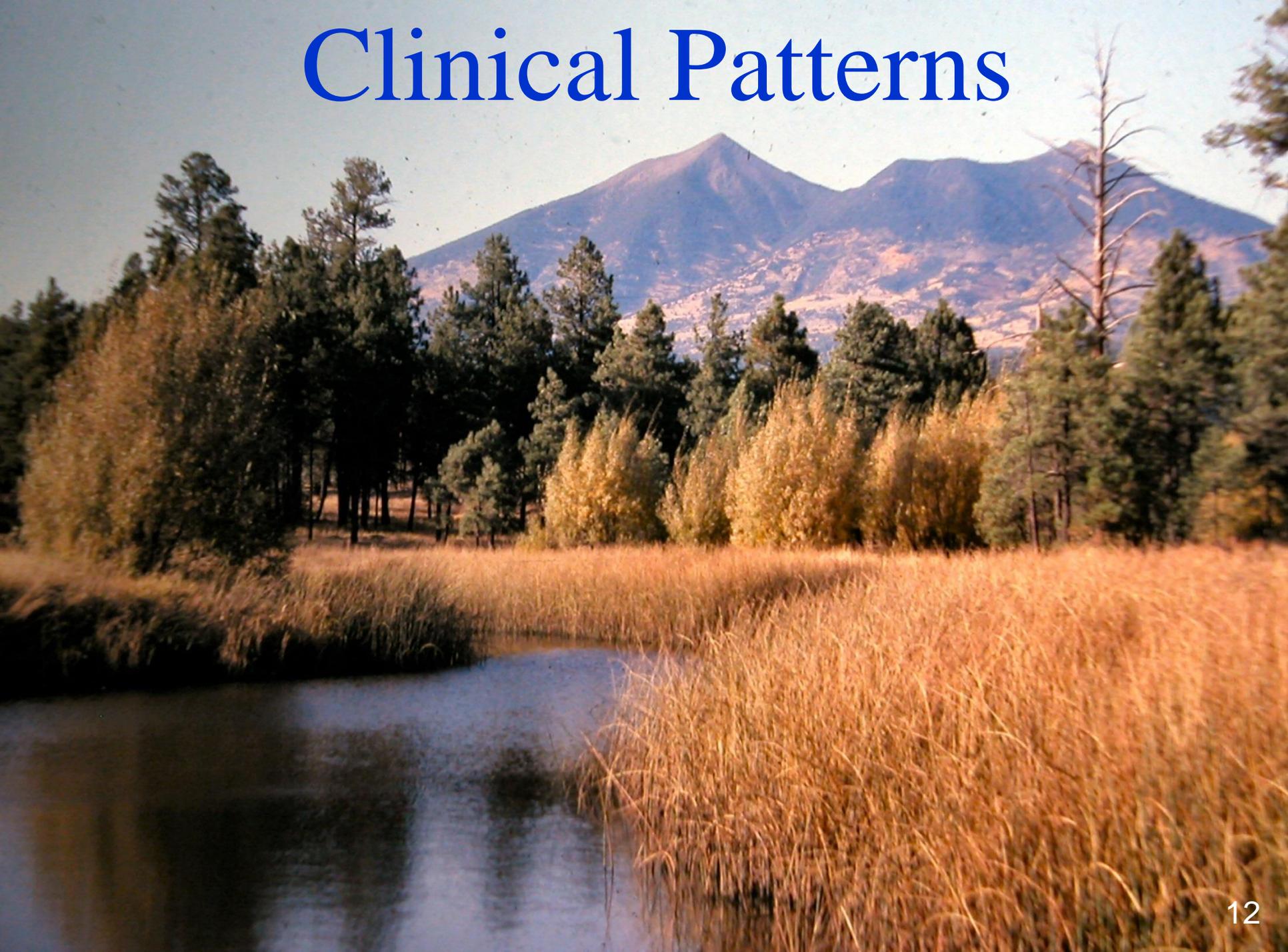


From Travis Am J Surg Pathol 2000 24(1):19-33

Evolution of Classification



Clinical Patterns



3 Clinical Patterns of ILD

1. Acute Diseases (hours/days to weeks)

- DAD (AIP), EP, Vasculitis/DAH, Drug, CVD

2. Subacute Diseases (weeks to months)

- HSP, Sarcoid, NSIP, Drugs, Smoking (RBILD and PLCH), “Chronic” EP, Constrictive bronchiolitis, CVD

3. Chronic Diseases (months to years)

- UIP, Fibrotic NSIP, Pneumoconioses, CVD-related
Chronic HSP, Smoking (DIP/RBILD and PLCH),
Constrictive bronchiolitis, angiopathic diseases

A dramatic landscape photograph capturing a sunset or sunrise. The sky is filled with dark, heavy clouds, with a bright, glowing area where the sun is setting or rising, creating a strong orange and yellow light that illuminates the lower part of the sky and the horizon. In the foreground, a silhouette of a windmill is visible against the bright horizon line. The overall mood is serene and atmospheric.

Radiological Patterns

Tomographic Lung Pathology

*A field guide to HRCT patterns of
diffuse lung disease*

Key Elements

Pattern of abnormalities

Distribution of abnormalities

Evolution over time (tempo)

General Concepts

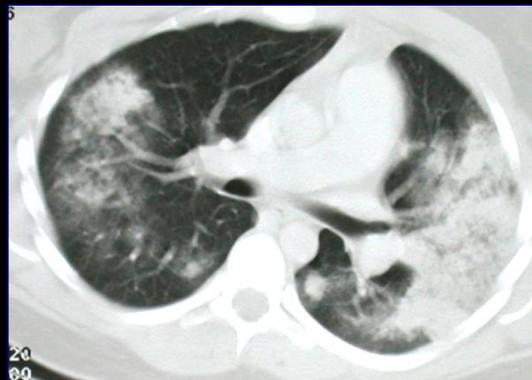
1. Upper lobe abnormalities= **INHALATION**
2. Lower lobe abnormalities= **VASCULAR**
3. Central abnormalities= **CARDIAC**
4. Peripheral abnormalities= **IMMUNOLOGIC**

HRCT- Patterns



- Ground Glass
- Consolidation
- Reticular and linear densities
- Nodular Opacities
- Mosaic pattern
- Cystic

The 4 CT Patterns of Pulmonary Disease



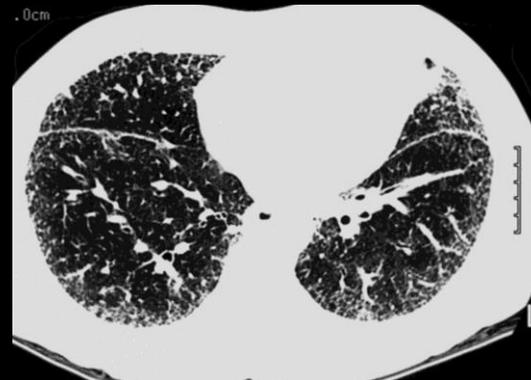
Ground Glass and Consolidation

GGO

Alveolar Filling
Neutrophils
Edema
Macrophages
Hemorrhage
Fibrin/protein
Fibroblasts

Consolidation

Infection
Neoplasms



Reticulation with Distortion

Without HC cysts

Peripheral
NSIP
CVD
Chr Drug
Chr HP
UIP (early)

With HC Cysts

UIP
CVD
Local Scar



Nodules

Neoplasms
Infections
Subacute HP
RB-ILD
PLCH

Sarcoidosis
Certain Drugs
Rheumatoid nodules
Focal OP



Mosaic patterns and cysts

PHT
PVOD
LAM

Constrictive Bronch
PLCH
Emphysema

Histopathologic Patterns



CT Patterns

Ground Glass and Consolidation

**Fibrosis/
honeycombing**

Nodules/masses

Airtrapping + cysts

Histopathology Patterns

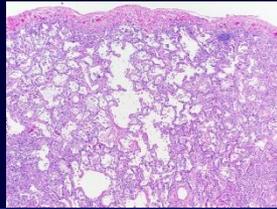
1. **Acute lung injury**
2. **Fibrosis**
3. **Cellular infiltrates**

4. **Alveolar filling**
5. **Nodules**
6. **Minimal changes**

The 6 Patterns of Interstitial Lung Disease

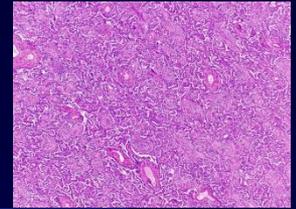
1. Acute Lung Injury

Prototype: DAD



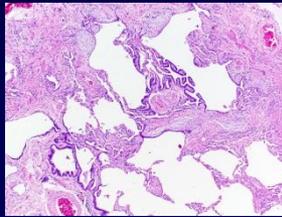
4. Alveolar Filling

Prototype: OP



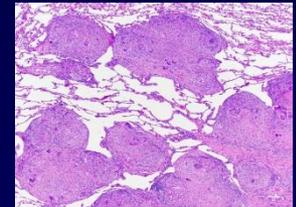
2. Fibrosis

Prototype:
UIP



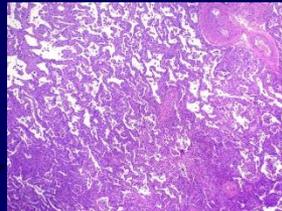
5. Nodules

Prototype:
Sarcoidosis



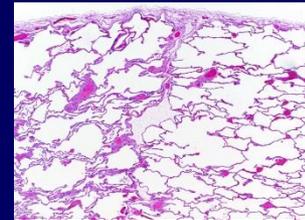
3. Cellular Infiltrates

Prototype:
Hypersensitivity



6. Minimal Changes

Prototype: SAD



The 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?

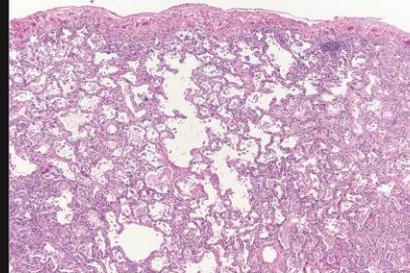
6. Minimal Changes

- with small airways disease?
- with vascular changes?
- with cysts?
- sampling problem?

The 6 PATTERNS of PULMONARY PATHOLOGY

Kevin O. Leslie, M.D.

1. Acute Lung Injury



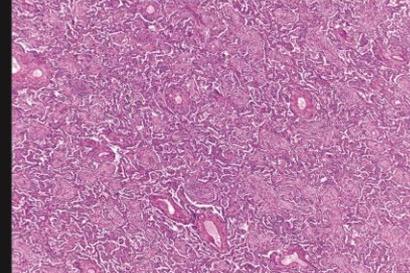
w/ Hyaline membranes (DAD)
Infection
Collagen vascular disease
Drug toxicity
Idiopathic acute interstitial pneumonia
Clinical ARDS

w/ Leukostasis
Acute nonthrombotic pneumonia
Drug toxicity
Diffuse alveolar damage in smokers

w/ Necrosis
Infectious
- Viral
- Bacterial
- Fungal
DAD in smokers
Some tumors

w/ Edema/hemorrhage
Diffuse alveolar hemorrhage
Collagen vascular diseases
DAD in smokers

4. Alveolar Filling



w/ Microthrombi
Smoking related
(RB, RHL, DTRP)
Local fibrosis effect
Airway obstruction

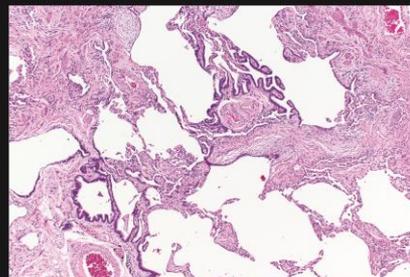
w/ Acute fibrin
Diffuse alveolar hemorrhage

w/ Organizing pneumonia
Infection
Drug toxicity
Collagen vascular diseases
Cryptogenic organizing pneumonia
(s.k.a. idiopathic BOOP)

w/ Extracellular material
Infection
Collagen vascular diseases
Drug toxicity
Diffuse alveolar hemorrhage
Pulmonary alveolar proteinosis

w/ Hemorrhage
Collagen vascular diseases
Drug toxicity
Diffuse alv. hemorrhage
Atrial
Congestive heart failure

2. Fibrosis



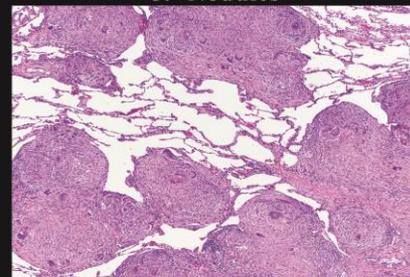
w/ Variable fibrosis (normal vs. honeycombing)
Usual interstitial pneumonia/HP
Adenocarcinoma
Extramural arthritis
Chronic hypersensitivity pneumonitis
Pulmonary Langerhans cell histiocytosis (mainly as sulfate scars)

w/ Honeycombing only
Diffuse alveolar opacities
Usual interstitial pneumonia (late)

w/ Diffuse alveolar wall fibrosis
Collagen vascular disease
Drug toxicity
Collagen vascular diseases

w/ Fibrosis
Collagen vascular diseases
Sarcoid (with granulomas)
Pneumoconiosis
Fibrotic nonspecific interstitial pneumonia (NSIPF)

5. Nodules



w/ Lymphoid cells
Follicular bronchiolitis
Wegener's
Diffuse lymphoid hyperplasia
Lymphoma

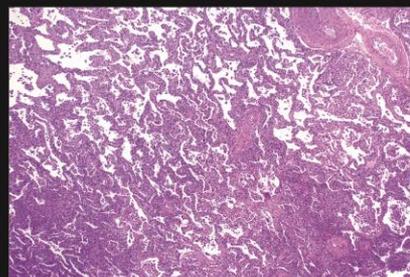
w/ Necrosis
Infectious
Neoplasms
Wegener's
Sarcoma

w/ Atypical cells
Infectious
Carcinoma
Lymphoma
Sarcoma

w/ Shallow scars
Pulmonary Langerhans cell histiocytosis (PLCH)

With Organizing Pneumonia
Organizing infections
Collagen vascular diseases
Drug toxicity
Wegener's
Organizing aspiration
Infarct

3. Cellular Infiltrates



w/ Lymphocytes and plasma cells
Hypersensitivity pneumonitis
Collagen vascular diseases
Lymphoid interstitial pneumonia
Nonspecific interstitial pneumonia, cellular form
Certain drug toxicities and infections
Lymphoproliferative diseases

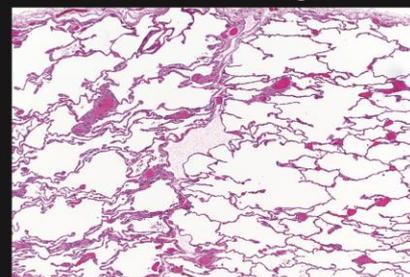
w/ Neutrophils
Infection
Collagen vascular diseases
Hemorrhage syndromes

w/ Granulomas
Infectious
Hypersensitivity pneumonitis
Sarcoidosis/BoS
Aspiration

w/ Focal organizing pneumonia
Infection
Collagen vascular diseases

w/ Fibrosis
Collagen vascular diseases

6. Minimal Changes



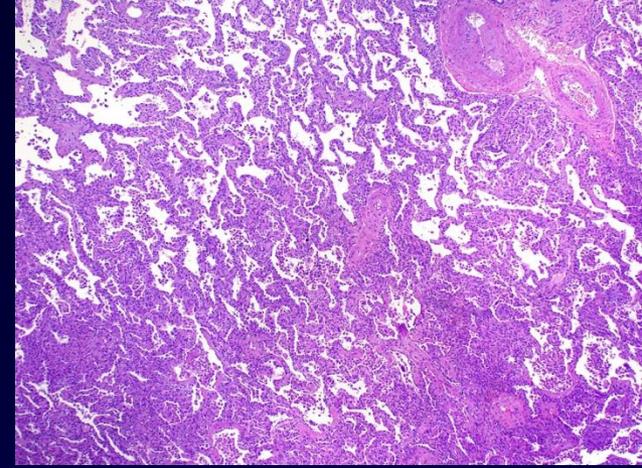
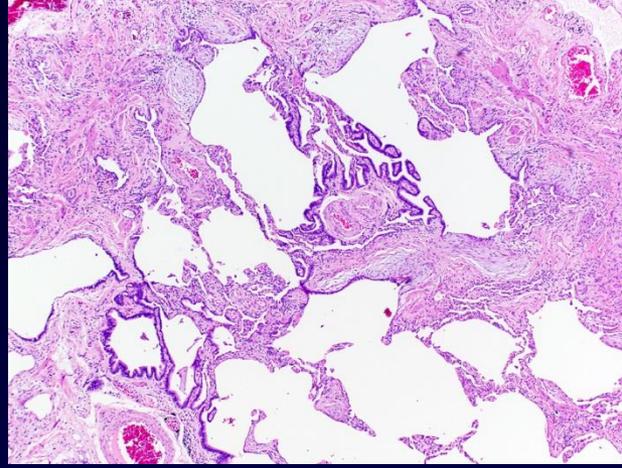
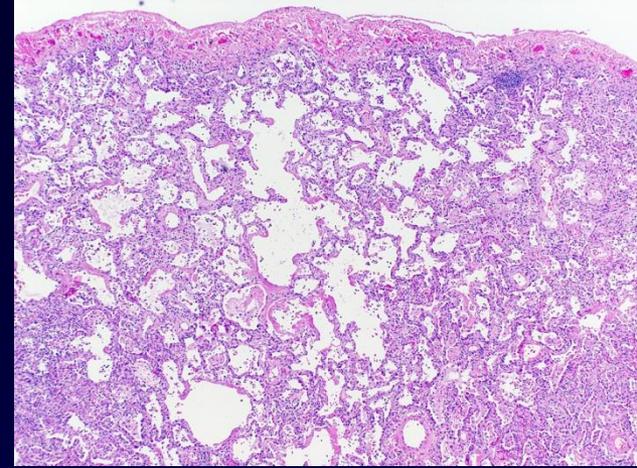
w/ Small Airway Disease
Constrictive bronchiolitis

w/ Vascular Disease
Pulmonary hypertension
Veno-occlusive disease (PVOD)

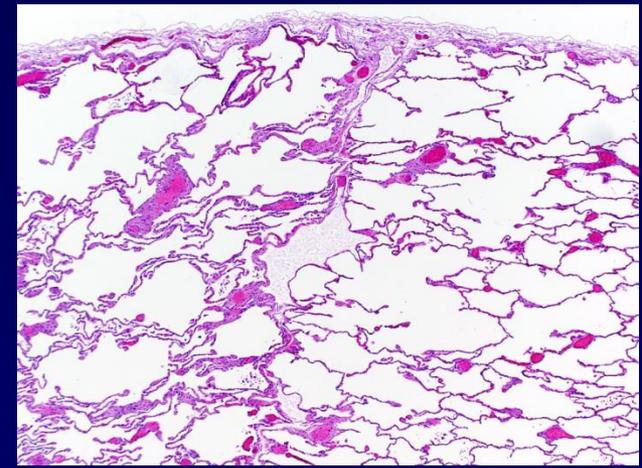
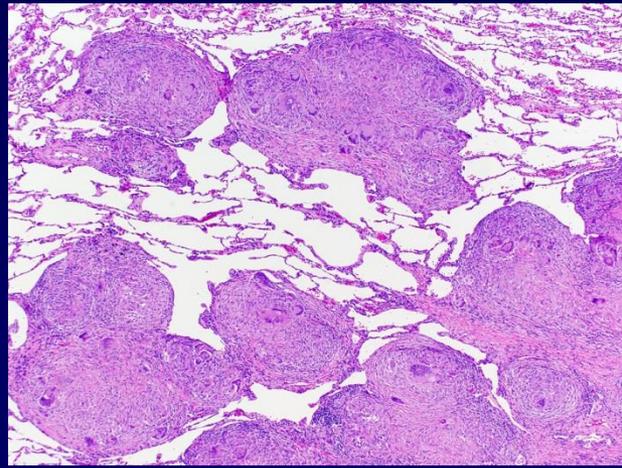
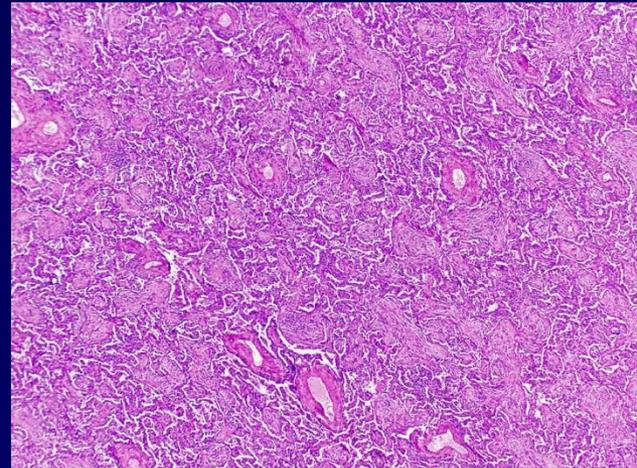
w/ Edema
Pulmonary Langerhans cell histiocytosis (PLCH)
Lymphangioleiomyomatosis (LAM)

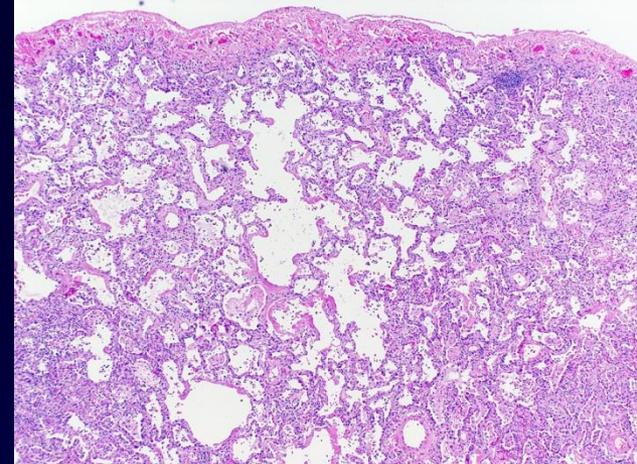
w/ No specific findings
Early acute injury?
Pulmonary edema?
Sampling error?

Differential diagnosis based on additional findings

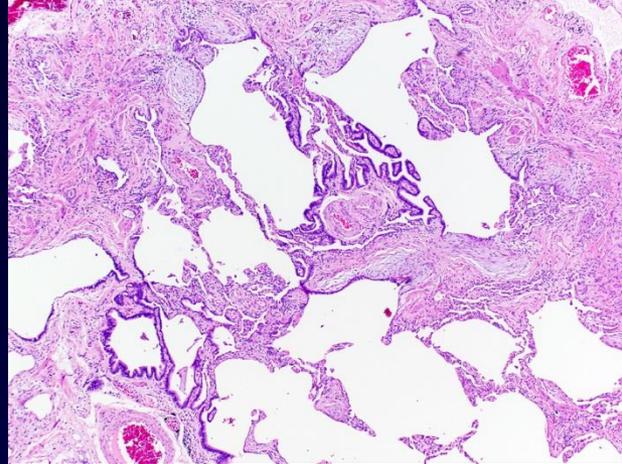


Post-Test

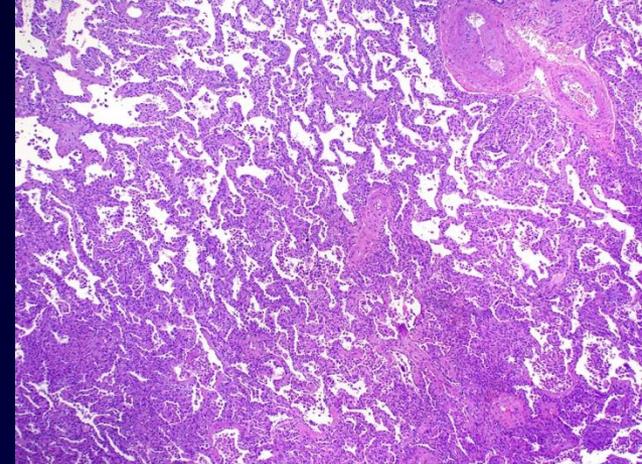




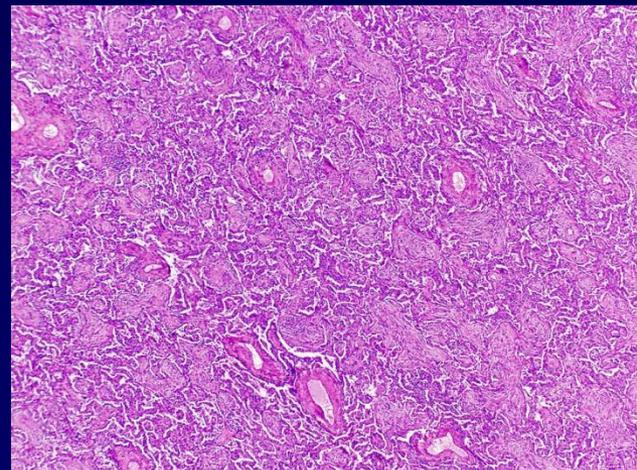
Diffuse Alveolar Damage



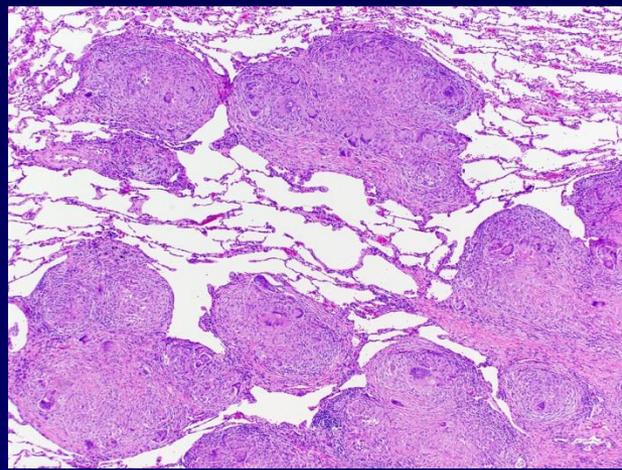
Usual Interstitial Pneumonia



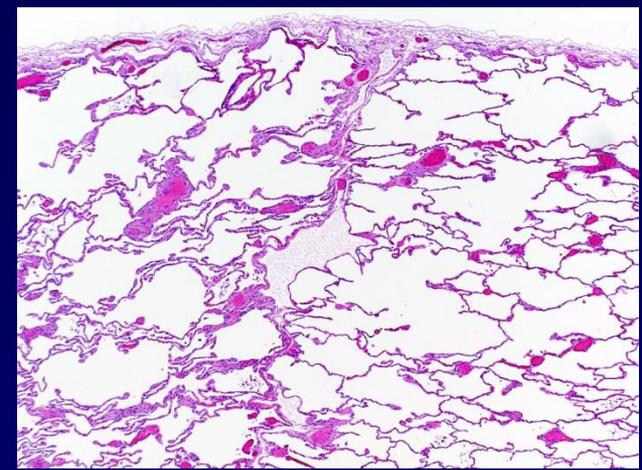
Hypersensitivity Pneum.



Organizing Pneumonia



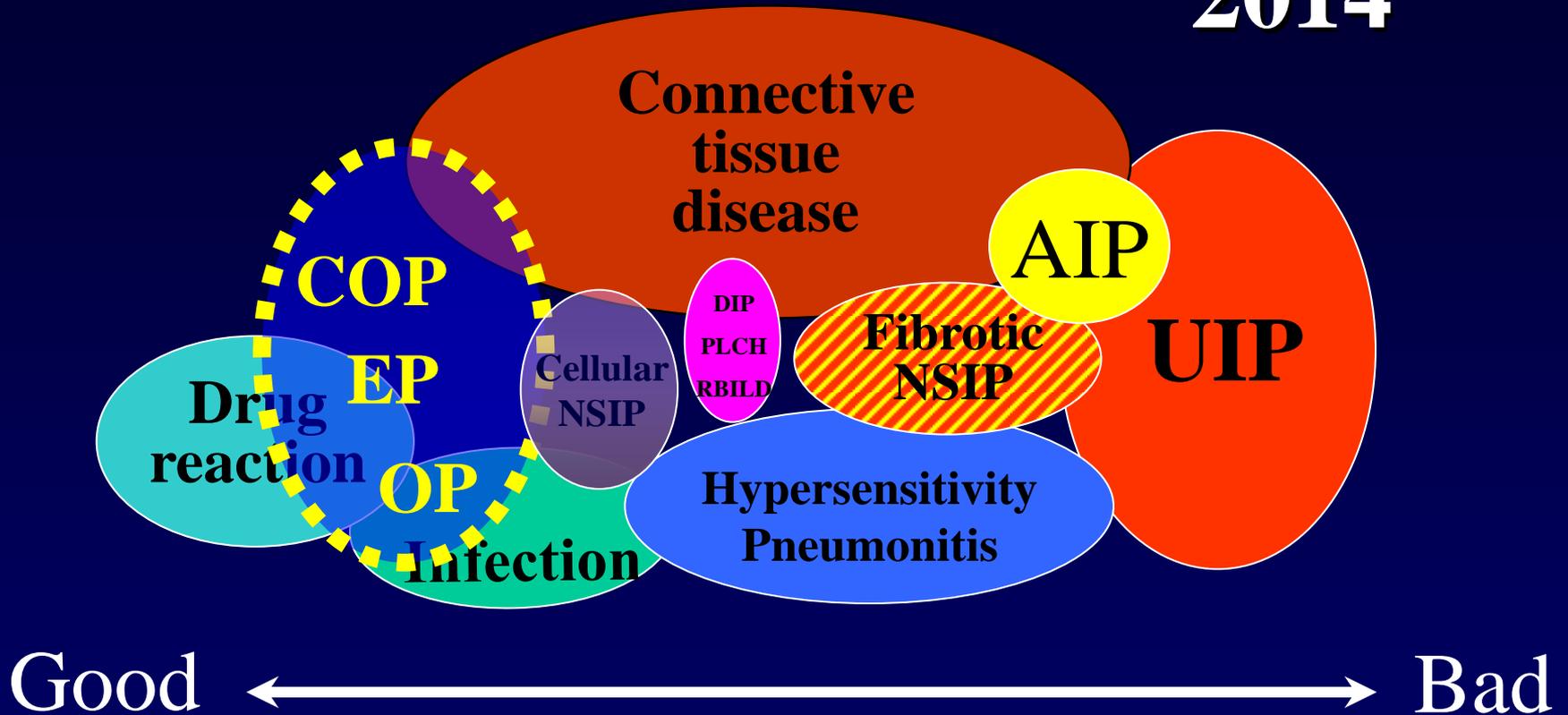
Sarcoidosis



Constrictive Bronchiolitis

Diagnosis Matters!

2014



Prognostic Classification of ILD

Interstitial Lung Disease

Clinical, Radiologic and Pathologic Patterns

Clinical Patterns (3) and Differential

Acute (Hrs/days): Infection, ARDS, EP, Drug and toxin reactions, Vasculitis, Hemorrhage, AIP, Acute exacerbation of chronic disease

Subacute (Weeks/Months): HP, Smoking, Sarcoid, Rheumatic, Drugs, Chronic (persistent / slowly resolving) pneumonias, BOOP/COP, NSIP, LIP, Chronic EP.

Chronic (Many months/years): Rheumatic, Drugs, Pneumoconiosis, Sarcoid, Smoking, Small airway disease, UIP, AP, Amyloid

Radiologic (HRCT) Patterns (4) and Differential:

Ground glass/ consolidation: Edema, infection, hemorrhage, organizing pneumonia, infarction, diffuse alveolar damage (any cause), delicate interstitial fibrosis (NSIP-like)

Reticulation and distortion: Fibrosis (any cause)

Nodules: Infection (often granulomatous), aspiration, pneumoconiosis, infarction, neoplasm, sarcoidosis, Wegener granulomatosis, Langerhans cell histiocytosis (PLCH)

Mosaic pattern/ cysts: Constrictive bronchiolitis, pulmonary vascular diseases, LAM, PLCH

The Rosetta Stone

Pathologic Patterns (6) and Differential

Acute lung injury: Infection, drug toxicity, autoimmune disease, idiopathic forms (AIP and AFOP), Acute on chronic

Fibrosis: UIP, NSIP, chronic drug toxicity, autoimmune diseases, rare genetic diseases, focal scars (any cause)

Cellular infiltrates: Hypersensitivity pneumonitis, drug toxicity, autoimmune diseases, certain chronic infections, NSIP, LIP, low grade lymphoma

Alveolar filling: Edema, acute pneumonia, organizing pneumonia (any cause), alveolar proteinosis, alveolar microlithiasis, dendriform calcification, aspiration

Nodules: Infection (often granulomatous), aspiration, pneumoconiosis, infarction, neoplasm, sarcoidosis, Wegener granulomatosis, Langerhans cell histiocytosis (PLCH)

Minimal changes: Constrictive bronchiolitis, pulmonary vascular diseases, LAM, PLCH



My approach to interstitial lung disease using clinical, radiological and histopathological patterns

K O Leslie

J. Clin. Pathol. 2009;62;387-401
doi:10.1136/jcp.2008.059782



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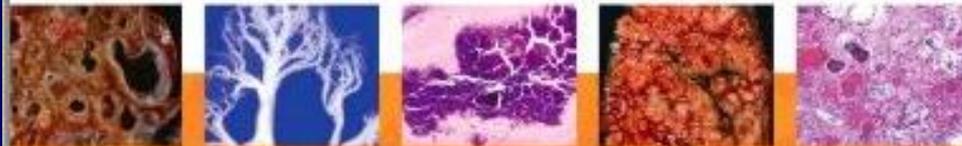
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Second Edition

PATTERN RECOGNITION SERIES
SERIES EDITORS: KEVIN O. LESLIE AND MARK R. WICK

Practical Pulmonary Pathology

A Diagnostic Approach



Kevin O. Leslie
Mark R. Wick

ELSEVIER
SALINDERS

C
O
P

Questions?

LIP

Eosinophilic pneumonia

Infection

Drugs

U

RBILD/DIP

Connective tissue diseases

Pneumoconioses

PLCH
PLCH

I

vasculitis

NSIP

P

Sarcoid

Hypersensitivity