SPECTRUM OF ILD IN CONNECTIVE TISSUE DISEASE

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DISCLOSURE STATEMENT

NO DISCLOSURE/ CONFLICT OF INTEREST

OUTLINES

4

3

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- INTRODUCTION TO CTD-ILD
- BASIC GLOSSARIES OF HRCT TERMINOLOGY
- PATTERN SUBTYPES OF CTD-ILD ON HRCT
- PROGNOSTIC FACTORS OF CTD-ILD ON HRCT

INTRODUCTION

INTRODUCTION

- Interstitial Lung Disease - A broad category of heterogeneous diffuse parenchymal lung disorders resulting from widespread inflammatory and/or fibrotic process.
- =Diffuse parenchymal lung disease
- >300 conditions fall in this category
- Only 1/3 has a known cause
- Survival rates at 5 years 20% for IPF and almost 100% for COP

Major ILDs of known aetiology (~35% of all patients with ILDs)

- Pneumoconioses (e.g. asbestosis, silicosis)
- Extrinsic allergic alveolitis (hypersensitivity pneumonitis)
- Iatrogenic ILD caused by drugs and/or radiation
- Post-infectious ILD

Major ILDs of unknown aetiology (~65% of all patients with ILDs)

- Sarcoidosis
- Idiopathic interstitial pneumonias, of which the most important are:
  - IPF with a histopathological pattern of usual interstitial pneumonia (~55% of IIPs)
  - Nonspecific interstitial pneumonia (~25% of IIPs)
  - Respiratory bronchiolitis ILD, occurring in smokers (~10% of IIPs)
  - Desquamative interstitial pneumonia (~5% of IIPs)
  - Cryptogenic organising pneumonia (~3% of IIPs)
  - Lymphoid interstitial pneumonia (~1% of IIPs)
  - Acute interstitial pneumonia (~1% of IIPs)
- ILD in CTDs and in collagen-vascular diseases, of which the most important are:
  - ILD in rheumatoid arthritis
  - ILD in progressive systemic sclerosis

Table 1 – Classification of interstitial lung diseases (ILDs). IIP: idiopathic interstitial pneumonia; IPF: idiopathic pulmonary fibrosis; CTDs: connective tissue diseases.

The European lung white book. ERS 2013

- ILD in CTD causes mortality and morbidity
- The patterns in CTD-ILD mirrors those seen in IIP
- Lung injury is not restricted to ILD-

  (iatrogenic /oportunistic infections, follicular bonrchiolitis/ diffuse bronchiectasis/ BO etc)

PREVALENCE/FREQUENCY OF ILD IN CTD

Interstitial lung diseases associated with connective tissue diseases

Rheumatic disease	Frequency of ILD (%)	Comment
Systemic sclerosis	45 (clinically significant)	More common in diffuse disease; topoisomerase-1 antibodies
Rheumatoid arthritis	20 to 30	Increased risk with cigarette smoking
Polymyositis/dermatomyositis	20 to 50[a]	More common with anti-synthetase antibodies
Sjögren's syndrome	Up to 25	-
Systemic lupus erythematosus	2 to 8	Usually in patients with multisystem disease
Mixed connective tissue disease	20 to 60	-

ILD, interstitial lung disease. [a]Frequency may be higher based on recent studies.

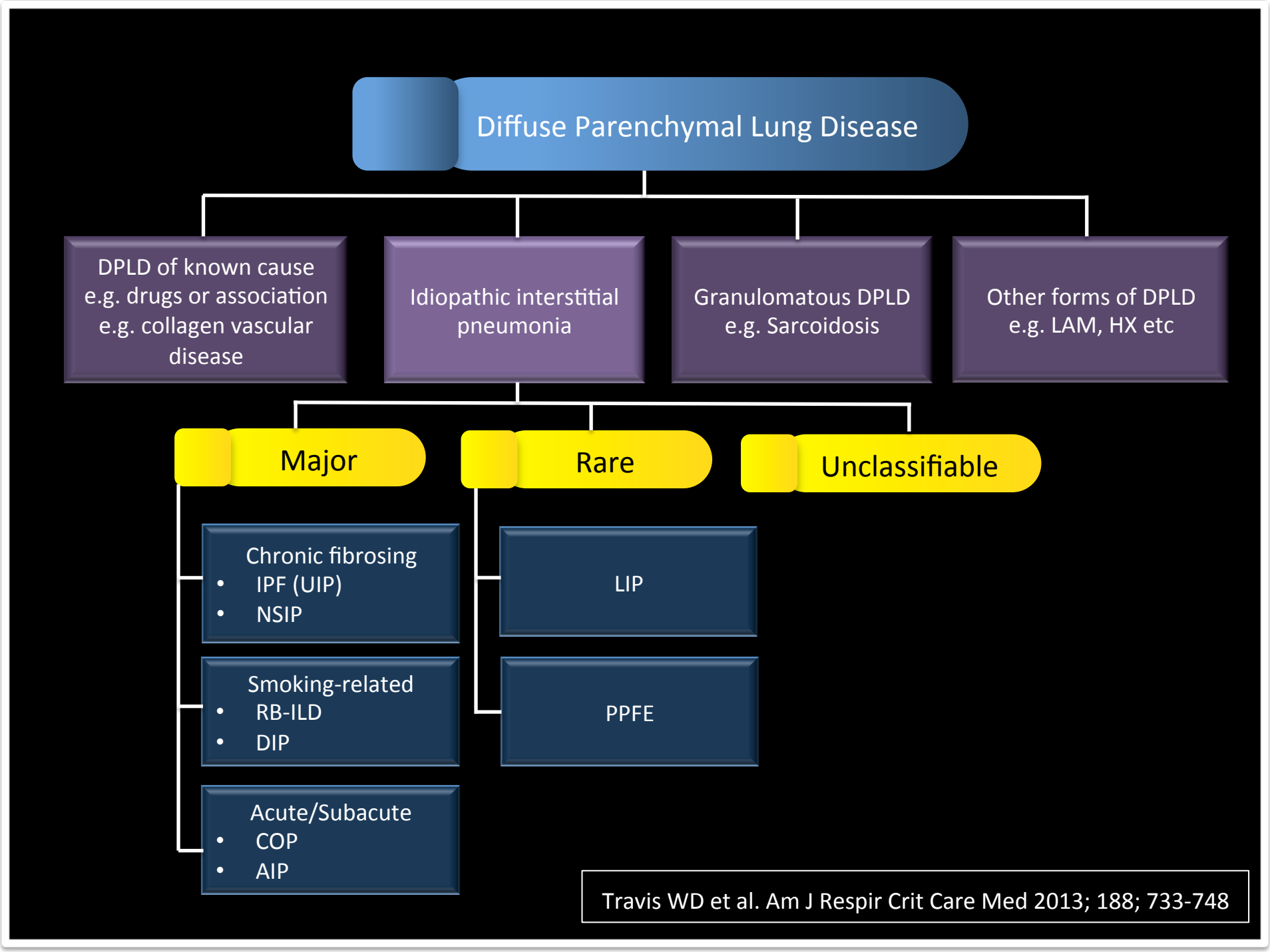
Castelino et al. *Arthritis Research & Therapy*. 2010;12(4):213.

	ILD	Airways	Pleural	Vascular	DAH
Systemic sclerosis	+++	–	–	+++	–
Rheumatoid arthritis	++	++	++	+	–
Primary Sjögren's syndrome	++	++	+	+	–
Mixed CTD	++	+	+	++	–
Polymyositis/ dermatomyositis	+++	–	–	+	–
Systemic lupus erythematosus	+	+	+++	+	++

The signs show prevalence of each manifestation (–=no prevalence; +=low prevalence; ++=medium prevalence; +++=high prevalence). ILD=interstitial lung disease. DAH=diffuse alveolar haemorrhage. CTD=connective tissue disease.

Table 1: **CTDs and common pulmonary manifestations**

Fischer et al. The Lancet. 2012;380:689-98.


Diffuse Parenchymal Lung Disease
DPLD of known cause e.g. drugs or association e.g. collagen vascular disease
Idiopathic interstitial pneumonia
Granulomatous DPLD e.g. Sarcoidosis
Other forms of DPLD e.g. LAM, HX etc
Major
Rare
Unclassifiable
Chronic fibrosing
• IPF (UIP)
• NSIP
Smoking-related
• RB-ILD
• DIP
Acute/Subacute
• COP
• AIP
LIP
PPFE
Travis WD et al. Am J Respir Crit Care Med 2013; 188; 733-748