

## Idiopathic interstitial lung diseases

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**Authors:** I. Santiago<sup>1</sup>, M. Armas<sup>2</sup>, C. Ruivo<sup>3</sup>, M. A. Portilha<sup>3</sup>, M. Ferreira<sup>1</sup>,  
F. Figueiredo<sup>1</sup>, F. Caseiro-Alves<sup>3</sup>; <sup>1</sup>Aveiro/PT, <sup>2</sup>Barcelona/ES,  
<sup>3</sup>Coimbra/PT  
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### LEARNING OBJECTIVES

To briefly discuss the physiopathology of the entities globally known as Idiopathic Interstitial Lung Diseases (IILD), as well as their clinical findings, therapeutic options and prognosis;

To describe and illustrate the imaging findings of IILD, as seen on High Resolution Computer Tomography (HRCT) with severity correlation, when relevant.

To define a strategy for their differential diagnosis based on clinical and HRCT findings, with histological correlation.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## Background

Background

## BACKGROUND

ILD are classified, according to the American Thoracic Society and European Respiratory Society, as one of seven possible entities:

Idiopathic Pulmonary Fibrosis (IPF)

Nonspecific interstitial pneumonia (NSIP)

Cryptogenic organizing pneumonia (COP)

Respiratory bronchiolitis - interstitial lung disease (RB-ILD)

Desquamative interstitial pneumonia (DIP)

Acute interstitial pneumonia (AIP)

Lymphoid interstitial pneumonia (LIP)

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## BACKGROUND

Their diagnosis is based on clinical, HRCT and histological findings except for IPF, for which clinical and HRCT findings suffice in most cases.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

**Imaging findings OR Procedure details**

## IMAGING FINDINGS

We present the typical HRCT imaging findings of all the above-mentioned entities, with clinical and histological correlation, based on selected cases from our Institution's database, emphasizing the imaging features that help us define a strategy for their differential diagnosis.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## UIP / IPF

- Usual interstitial Pneumonia (UIP) refers to the histologic pattern, whereas Idiopathic Pulmonary Fibrosis (IPF) refers to the clinical entity
- ♀ and ♂ equally affected
- Age at presentation usually > 50 years
- Clinical findings (mostly dyspnea on exertion) with 3 or more months duration
- Diagnosis based on clinical, functional and HRCT findings only, unlike all other IILD, for which lung biopsy is required
- Treatment is largely ineffective and prognosis is very poor (mean survival 3 to 4 years)

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



## UIP / IPF

### HRCT findings

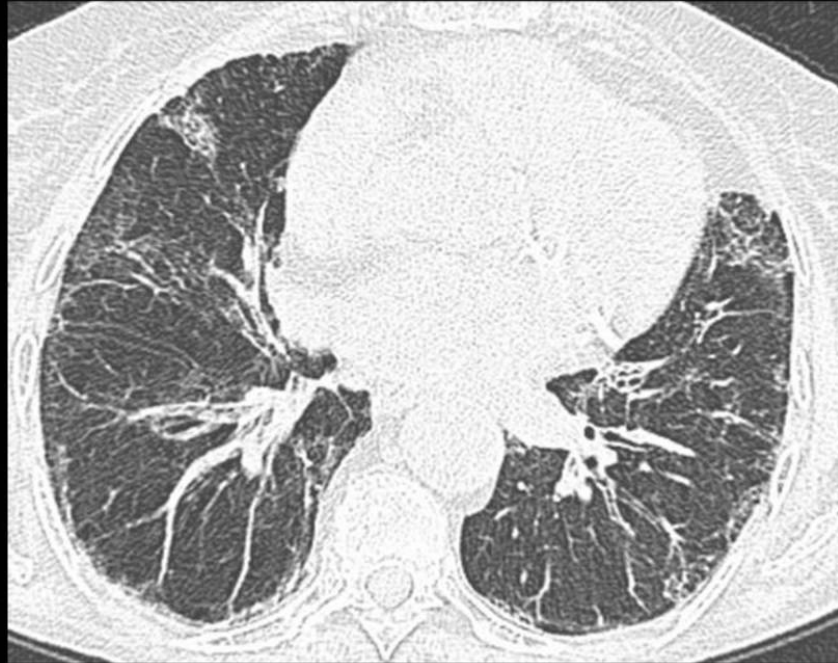
- Irregular reticular opacities
- Architectural distortion and volume loss
- Traction bronchiectasis
- Areas of ground-glass attenuation (not a prominent feature)
- Honeycombing
- Mediastinal lymph node enlargement

### Distribution

- Predominantly basal and peripheral
- Usually simetric

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

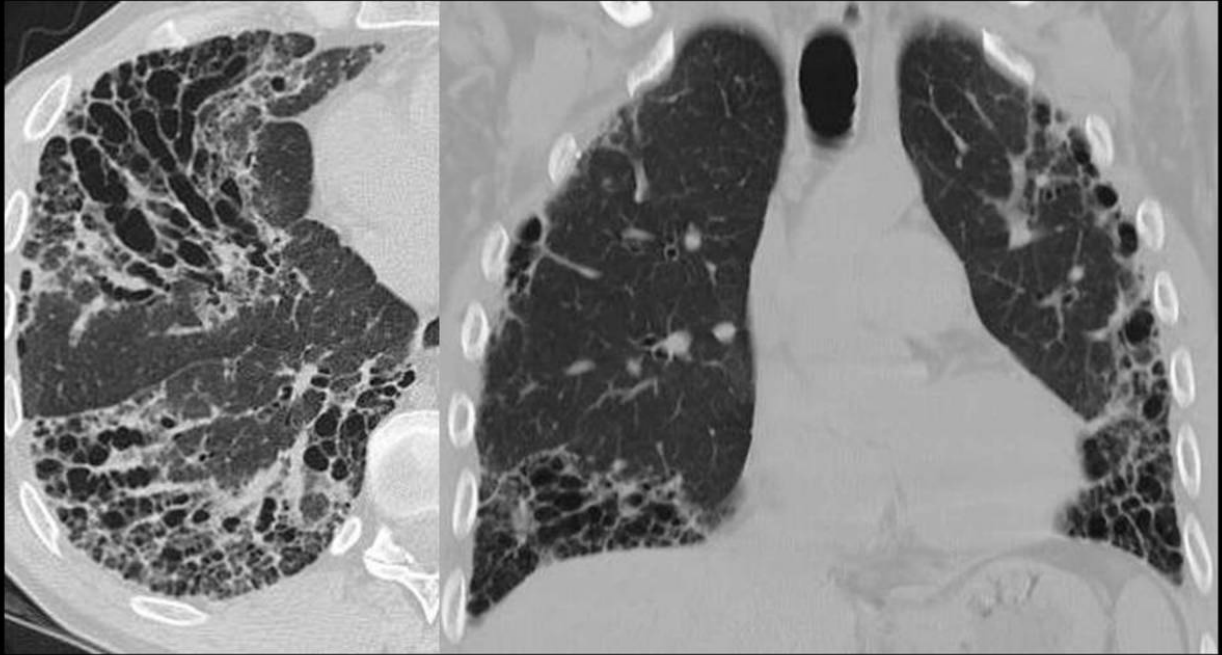


**Figure 1 UIP/IPF** Early findings consisting of fine reticular opacities with a predominantly basal and peripheral distribution.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL





**Figure 2 UIP/IPF** Late findings, consisting of coarse reticular opacities, honeycombing and traction bronchiectasis.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## NSIP

- Considered by some as a diagnosis of exclusion and by others a specific entity
- Histologically, temporal homogeneity of the inflammation and fibrosis distinguishes it from UIP/IPF
- 2 distinct histologic patterns may be found: cellular and fibrotic
- ♀ and ♂ equally affected
- Age at presentation usually between 40 and 50 years
- Associated with collagen-vascular diseases, infection, drugs and hypersensitivity pneumonitis
- Steroid treatment is effective in most cases and prognosis is much better than UIP/IPF

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## NSIP

### HRCT findings

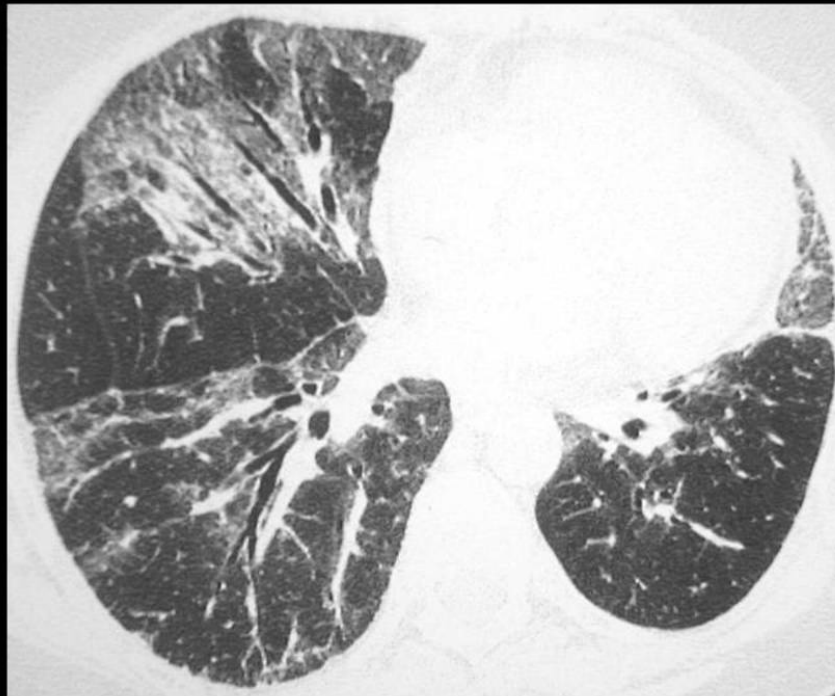
- Ground-glass opacification predominate in the cellular pattern)
- Irregular reticular opacities predominate in the fibrotic pattern. Honeycombing may also be found in this pattern but is uncommon and inconspicuous
- Patchy consolidation may also be found

### Distribution

- Bilateral, sometimes patchy
- Predominantly basal and peripheral with subpleural sparing

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 3 NSIP** Patchy ground-glass areas of attenuation and fine reticulation are seen bilaterally, with a predominantly basal and peripheral distribution.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 4 NSIP** Patchy ground-glass areas of attenuation with a predominantly basal and peripheral distribution. Note immediate subpleural sparing.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



## COP

- Idiopathic form of organizing pneumonia (which can be seen in association with infection, obstruction, aspiration, drugs, collagen-vascular diseases and others)
- ♀ and ♂ equally affected
- Mean age at presentation 55 years
- Symptomatology is similar to UIP/IPF but lasts less (may mimic infection) and systemic symptoms are more frequent
- Response to steroids is frequent and prognosis is good, although relapse is frequent

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



## COP

### HRCT findings

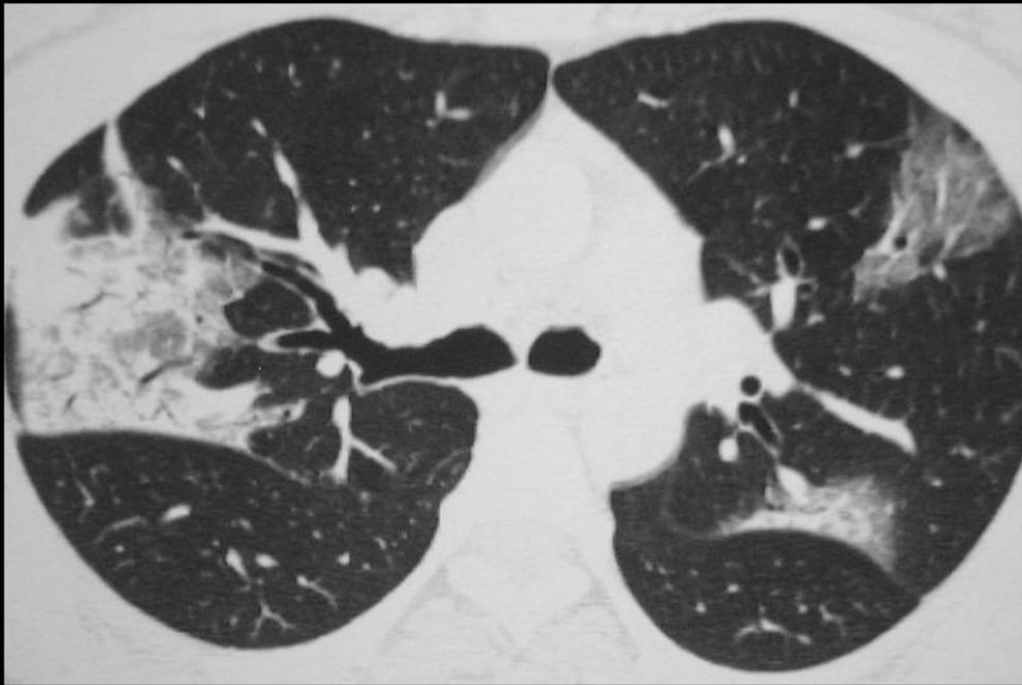
- Patchy consolidation
- Ground-glass opacification
- Centrilobular ill-defined nodules
- Large nodules or masses
- The atol sign

### Distribution

- Subpleural, peribronchial
- Lower lung zones more commonly affected
- Findings may be migratory

**Fig.:** \_

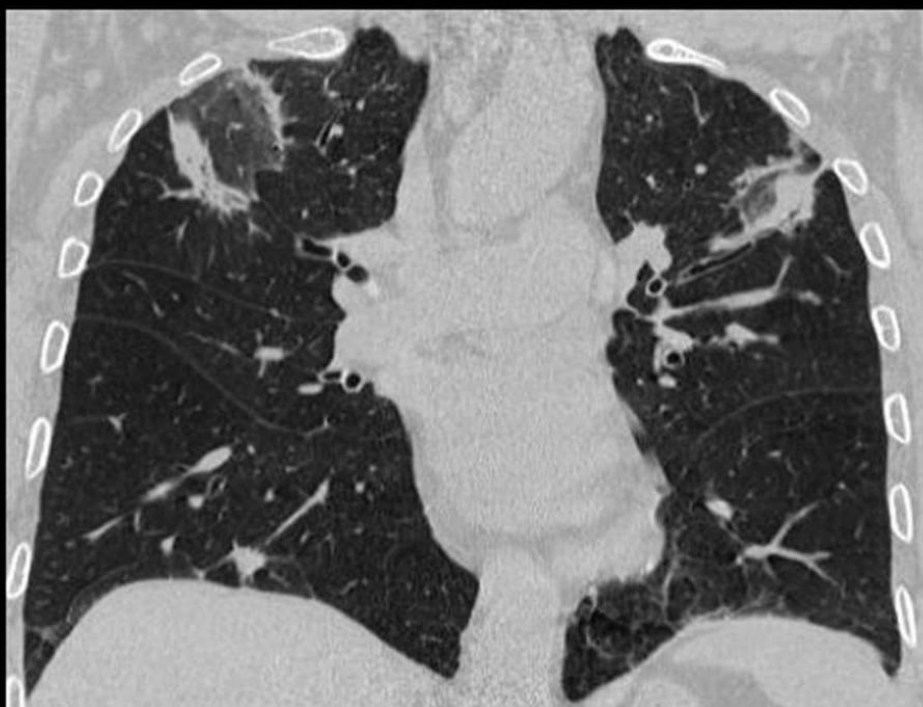
**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 5 COP** Patchy areas of consolidation and ground glass attenuation are found bilaterally, in a predominantly subpleural location.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 7 COP** The atol sign is depicted bilaterally. It consists of a circular/ring shaped area of consolidation surrounding a central area of ground-glass opacification.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## RB - ILD

- Symptomatic respiratory bronchiolitis seen almost exclusively in smokers
- ♂ are more commonly affected (2:1)
- Age at presentation between 30 and 40 anos
- Symptoms consist of slowly progressive dyspnea and dry cough

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## RB - ILD

### HRCT findings

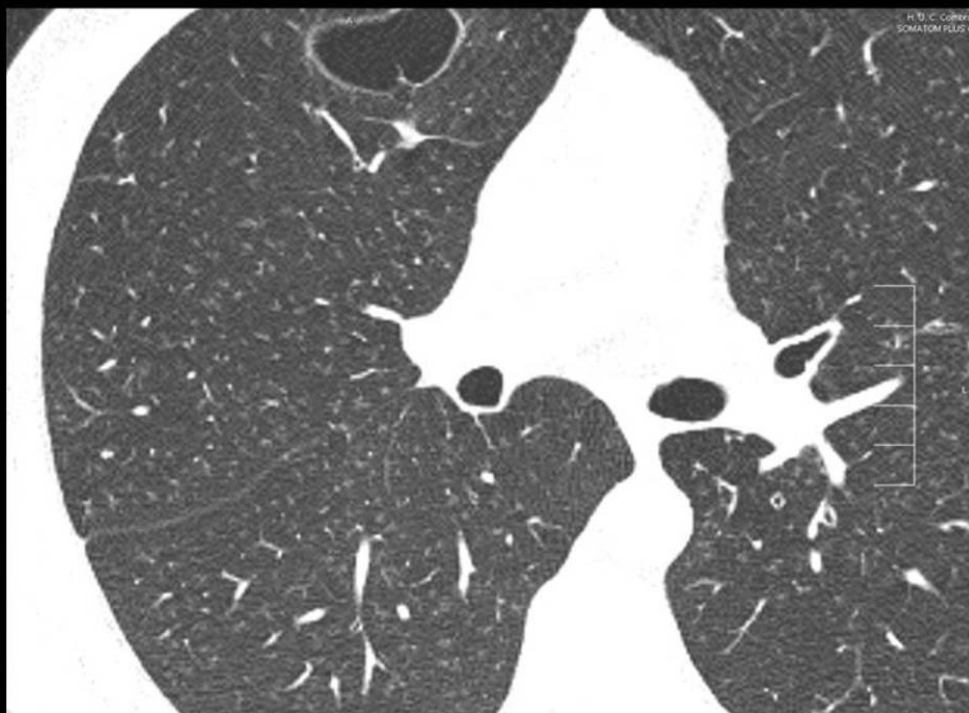
- Ground-glass opacification
- Centrilobular ill-defined nodules
- Thickening of bronchial walls
- Air-trapping
- Frequent coexistence of centrilobular emphysema

### Distribution

- patchy distribution of ground-glass areas of opacification
- Upper lobe predominance

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 8 RB-ILD** Ill-defined centrilobular nodules are depicted bilaterally. Note bullae in the anterior segment of the right upper lobe.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL





**Figure 9 RB-ILD** Ill-defined areas of ground-glass opacification with a patchy distribution.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 10 RB-ILD** Expiratory scan image depicting two areas of air trapping.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## DIP

- Related to smoking in over 90% of cases, whether active or passive (less common). Other rather infrequent associations include toxic inhalation, drugs, Langerhans cell histiocytosis, leukemia, asbestosis and pneumoconiosis
- Considered by some as an extreme form of presentation of RB-ILD
- ♂ more commonly affected (2:1)
- Age at presentation between 30 and 40 years
- Prognosis is good upon quitting smoking. Steroid therapy may also lead to significant improvement

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## DIP

### HRCT findings

- Ground-glass opacification
- Linear and irregular reticulation
- Small cysts
- Patchy air-trapping
- Frequent coexistence of centrilobular emphysema

### Distribution

- Predominantly basal and peripheral

**Fig.:** \_

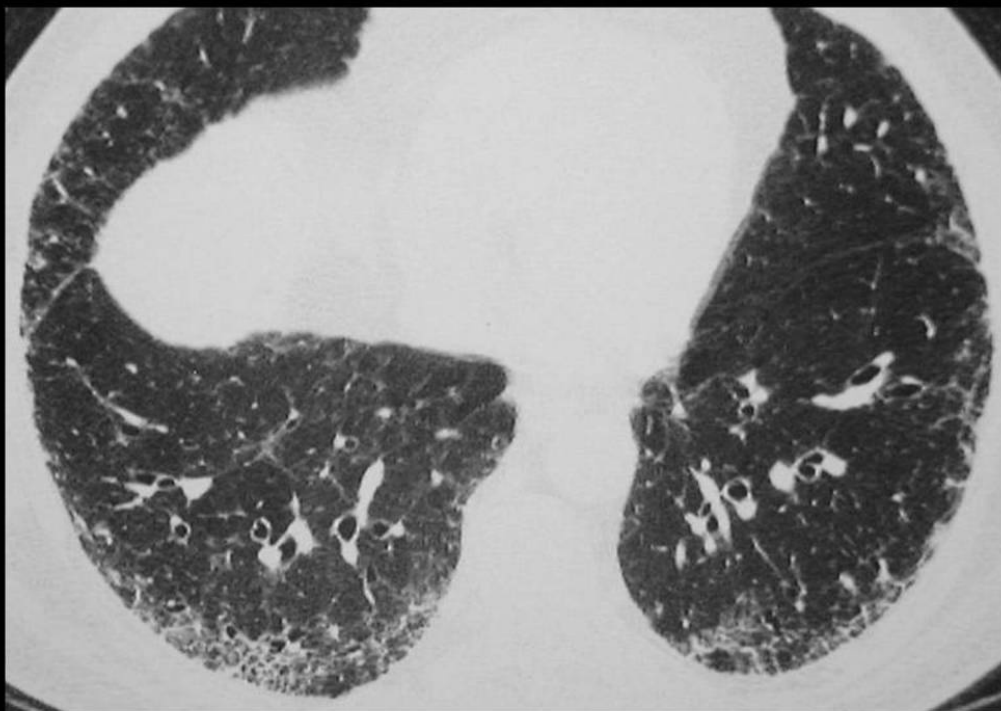
**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 11 DIP** Diffusely distributed areas of ground-glass opacification, these seen in both lower lobes, middle lobe and lingula. A cyst in the lingula is also seen.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



**Figure 12 DIP** Fine reticulation with a basal and peripheral distribution is the dominant feature in this patient.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



## AIP

- Fulminant idiopathic disease associated with the histologic pattern of diffuse alveolar damage, which may also be seen in acute respiratory distress syndrome
- Mean age at presentation is 50 years
- Symptoms similar to an upper respiratory infection may precede the disease
- Severe dyspnea rapidly followed by respiratory insufficiency represent the classical clinical picture
- Prognosis is bad with a mean survival of 50% at 6 months

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## AIP

### HRCT findings

- Ground-glass opacification
- Areas of consolidation
- Bronchial dilatation
- Reticular opacities, traction bronchiectasis, honeycombing and architectural distortion in late stages

### Distribution

- Diffuse, predominately basal or apical
- Ground-glass opacities and consolidation tend to be patchy
- Abnormalities tend to be more severe in the posterior zones

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## LIP

- Considered by some as a lymphoproliferative disease
- Rarely idiopathic. Frequently seen in association with collagen-vascular diseases, particularly Sjogren syndrome, but also with immunologic, infectious and drug related diseases
- ♂ more frequently affected than ♀
- Mean age at presentation 50 years
- Clinical findings other than the related disease include cough and dyspnea
- May respond to steroids but one third of patients progress to fibrosis

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

# LIP

## HRCT findings

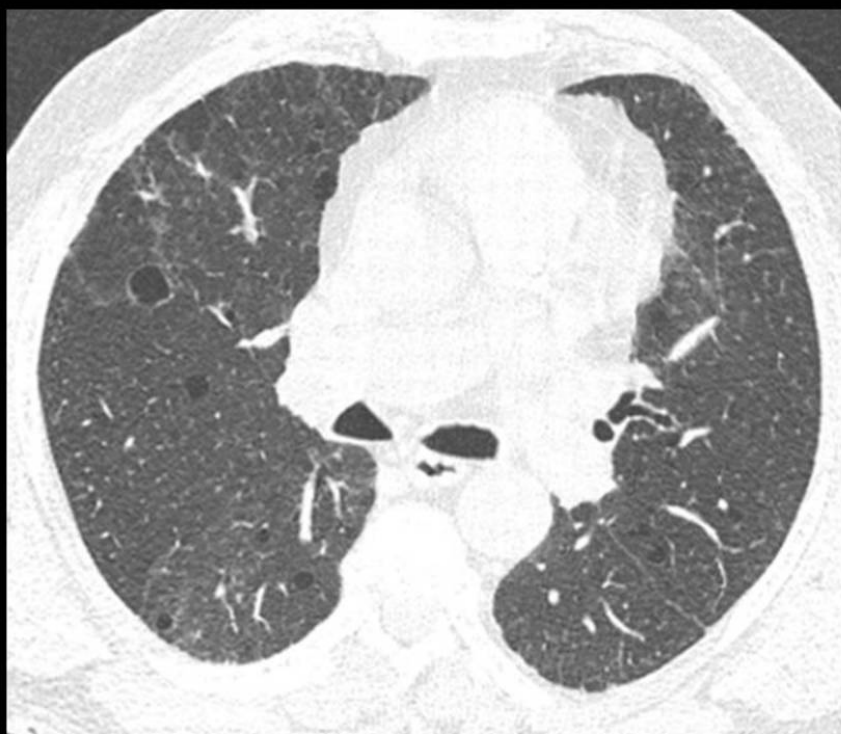
- Areas of ground-glass attenuation
- Areas of consolidation
- Ill-defined centrilobular nodules
- well-defined perilymphatic nodules and septal thickening
- Isolated perivascular cysts
- Diffuse cystic lesions mimicking honeycombing

## Distribution

- Predominantly basal

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

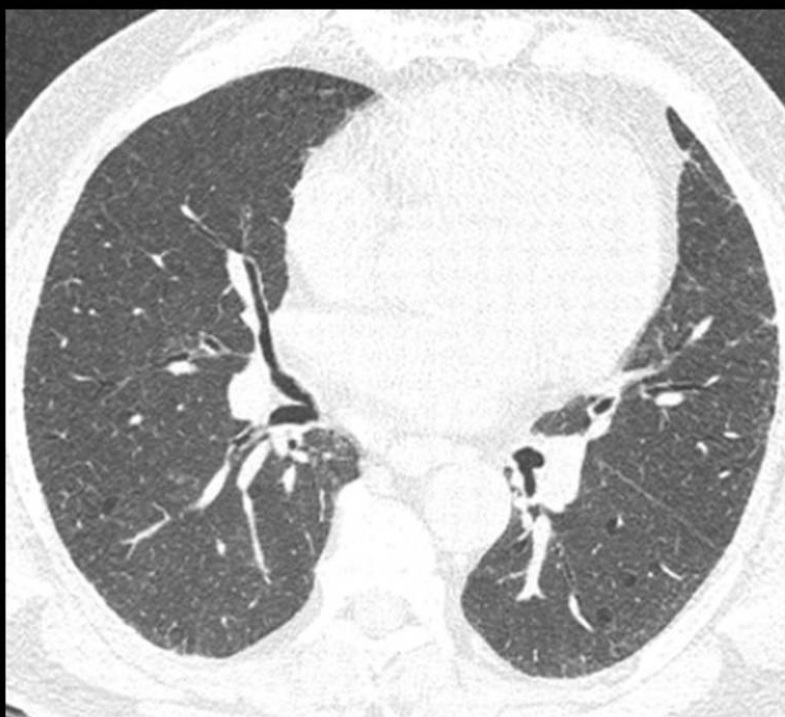


**Figure 13 LIP** Diffuse perivascular cysts associated with a few patchy areas of ground-glass attenuation and fine reticulation.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL



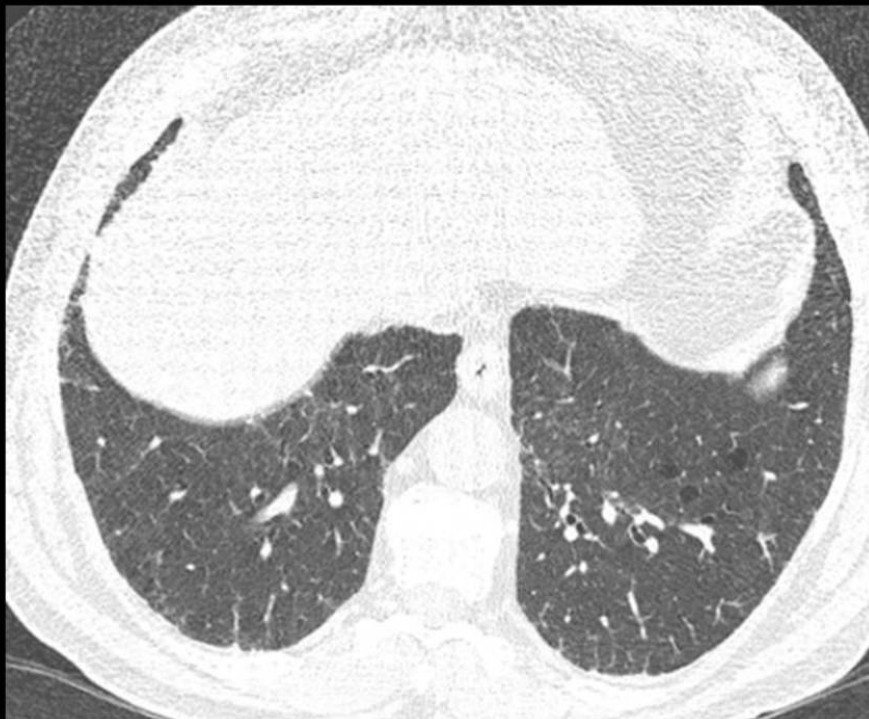


**Figure 14 LIP** Diffuse perivascular cysts associated with a few patchy areas of ground-glass attenuation and fine reticulation.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL





**Figure 14 LIP** Diffuse perivascular cysts associated with a few patchy areas of ground-glass attenuation and fine reticulation.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## TAKE HOME POINTS

- ❖ Acute Presentation is seen in AIP and COP
- ❖ Honeycombing is typical of UIP/FPI
- ❖ RB-ILD and DIP are symptomatic smoking-related diseases which respond to quitting smoking
- ❖ LIP is very rare
- ❖ Contrilobular ground-glass nodules with an upper lobe predominance suggest RB-ILD
- ❖ Thin walled cysts may be found in DIP and LIP
- ❖ Ground-glass opacification with sparing of immediate subpleural lung suggests NSIP

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## Conclusion

## CONCLUSIONS

The diagnosis of IILD is based on clinical, radiological and pathological grounds. HRCT is a very important tool for the diagnosis of IILD, even obviating the need for histological characterization when a usual interstitial pneumonia pattern is found, in the appropriate clinical setting.

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

## Personal Information

## AUTHOR INFORMATION

Inês Santiago

Serviço de Radiologia do Hospital Infante D. Pedro  
Aveiro, Portugal

inês\_agp\_santiago@hotmail.com

**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL

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**Fig.:** \_

**References:** I. Santiago; Radiology, Hospital Infante D Pedro - Aveiro, Aveiro, PORTUGAL