An Overview of Interstitial Lung Disease Imaging Reporting and Data System (ILD-RADS)

Fatma Zaiton[1], Al Shaimaa Fathi Elshetry[1], Eman El-Sayed Abdel Aziz, Ahmed Mohamed Alsowey[1]

Department of Radiodiagnosis, Faculty of Medicine Zagazig University. Egypt

Email: emyahmed14123@gmail.com

Abstract

Background: Interstitial lung diseases (ILDs) represent a large group of various lung diseases that cause pulmonary fibrosis. ILDs may cause diagnostic dilemmas for clinicians in daily practice due to numerous classifications and overlapping symptoms and imaging patterns. High-resolution computed tomography (HRCT) is the standard imaging method in the evaluation of ILDs. The recently introduced Interstitial Lung Disease Imaging Reporting and Data System (ILD-RADS) provide a standardized reporting template and four categories for the interpterion of ILDs patterns at HRCT.

This review focuses on the HRCT pulmonary features of ILDs and illustrates the ILD-RADS categories.

Keywords: Interstitial Lung Disease; HRCT; Interstitial Lung Disease Imaging Reporting and Data System (ILD-RADS).

Introduction

Interstitial lung diseases (ILDs) represent a large group of various lung diseases that cause pulmonary fibrosis with an increased risk of pulmonary failure and mortality **[1-2]**. ILDs can be mainly classified into ILDs with and without known underlying etiology **[3]**. Accurate diagnosis of ILDs is mandatory for optimal patient care **[4]**.

Several tools are used for diagnosing ILDs including detailed clinical history, physical examination, High-resolution computed tomography (HRCT), serological tests, lung function tests, bronchoalveolar lavage, and histo-pathological examination of lung biopsy **[5]**. Yet, diagnosis of ILDs is often difficult due to the overlap of their clinical, radiological, and histo-pathological features, and a multidisciplinary diagnostic approach integrating the clinical, imaging, and histopathological data is often required to establish a definitive diagnosis **[6]**.

High-resolution computed tomography (HRCT) of the chest is the standard imaging method in the evaluation of ILDs **[7]**. Four HRCT patterns of ILDs are described in the literature, including typical usual interstitial pneumonia (UIP), probable UIP, indeterminate for UIP, and inconsistent with UIP **[2]**. Accurate diagnosis of ILDs at HRCT requires careful interpretation of the imaging features and patterns **[8]**.

Berkowitz et al. recently introduced the Interstitial Lung Disease Imaging Reporting and Data System (ILD-RADS) to provide a standardized approach to report and categorize ILDs at HRCT into four ILD-RADS categories: ILD-RADS-1 (typical UIP); ILD-RADS-2 (possible UIP); ILD-RADS-3 (indeterminate for UIP); and ILD-RADS-4 (inconsistent with UIP) **[9].**

The final ILD-RADS category is assigned after the evaluation of specific pulmonary features and extrapulmonary findings at HRCT. Reporting of the pulmonary features includes the estimation of lung volume (normal, hypoinflated, or hyperinflated) as well as evaluating the presence or absence of pulmonary reticulations, traction bronchiectasis, honeycombing, pulmonary nodules, cysts, ground glass opacities (GGO), consolidation, mosaic attenuation, and emphysema. The axial (central, peripheral, and diffuse), and zonal distributions (upper, middle, and lower zones) of the pulmonary features should also be reported. Pulmonary complications such as acute infections, acute exacerbation, or cancer lung should be reported if present **[9].** Reporting of the extrapulmonary findings includes evaluating mediastinal, pleural,

tracheal, and bone or soft tissue lesions **[9]**. This review focuses on the pulmonary features of ILDs at HRCT and illustrates the ILD-RADS categories.

Pulmonary features of ILDs

1- Lung volume

Estimating lung volume, either normal, hypoinflated **(Figure 1),** or hyperinflated is best on the coronal or sagittal reformatted HRCT images **[1]**

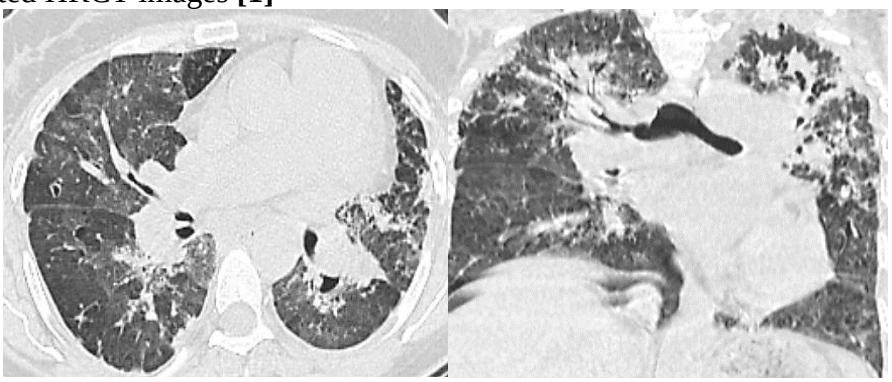

Figure 1: Axial and coronal HRCT images showing hypoinflated left lung with reticular and mosaic appearance.

2- Pulmonary reticulations

Pulmonary reticulations **(Figure 2)** refer to interstitial septal thickening, either smooth or nodular thickening. Pulmonary reticulations are often seen in association with traction bronchiectasis and honeycombing in UIP, connective tissue disease-ILD, and radiation-induced ILD **[10].**

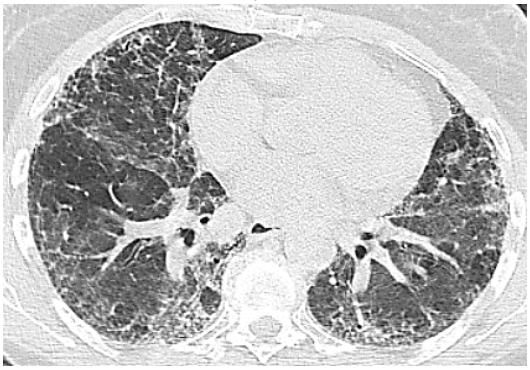

Figure 2: Pulmonary reticulations: axial HRCT image showing diffuse reticulations.

3-Traction bronchiectasis

Traction bronchiectasis **(Figure 3)** is an irregular and irreversible bronchial dilatation due to surrounding pulmonary fibrosis. Traction bronchiectasis is associated with pulmonary reticulations, GGO, and sometimes honeycombing **[11].**

Differentiating traction bronchiectasis from honeycombing on HRCT can be difficult. However, careful examination of consecutive thin-section, coronal, and sagittal reformatted CT images can help differentiate honeycomb cysts from branching traction bronchiectasis **[12-13].**

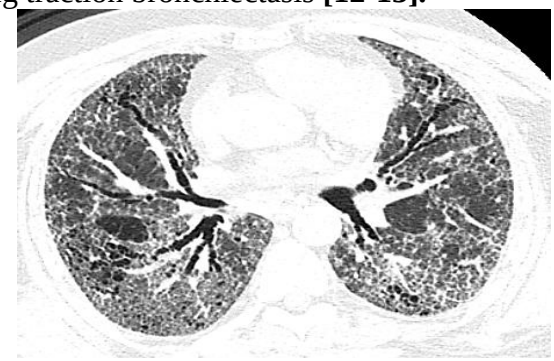

Figure 3: Traction bronchiectasis: axial HRCT image showing bilateral traction bronchiectasis.

4- Pulmonary honeycombing

Honeycombing **(Figure 4)** represents multiple small (3-10 mm in size) peripheral air-filled thin-walled cysts. Honeycombing is a characteristic sign of UIP, present in predominant basal and subpleural distributions **[14-15]**. However, minimal honeycombing with predominant GGO can be seen in non-specific interstitial pneumonia (NSIP) **[16]**. Honeycomb cysts have complete walls and are multilayered, these characteristics help differentiate honeycombing from emphysema and traction bronchiectasis **[13]**.

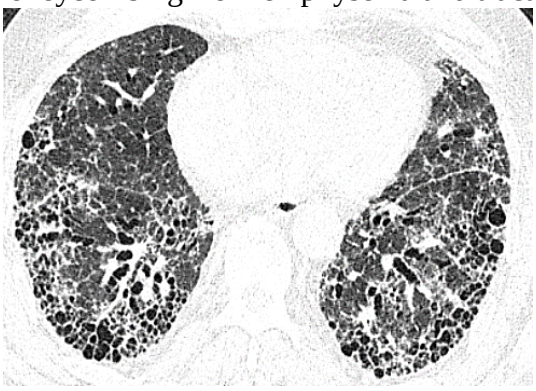

Figure 4: Pulmonary honeycombing: axial HRCT image showing bilateral honeycomb cysts in basal and subpleural predominance.

5- Pulmonary nodules

Pulmonary nodules **(Figure 5)** are multiple small (1mm-1cm in size) rounded opacities with smooth or irregular borders **[17-18]**. Pulmonary nodules can be classified into airspace nodules or interstitial nodules according to their origin. According to their distribution, pulmonary nodules can be classified into perilymphatic, centrilobular nodules, or miliary nodules. The presence of micronodules suggests an ILD other than IPF **[13]**. Perilymphatic nodules are typically in present sarcoidosis or silicosis seen subpleural, along the interstitial septa, fissures, and broncho-vascular bundles. Centrilobular nodules are seen in hypersensitivity pneumonitis (HP), separated from the pleura and interlobular septa **[17-18]**.

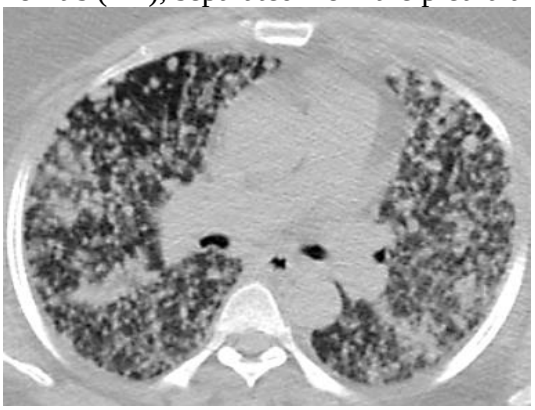

Figure 5: Pulmonary nodules: axial HRCT image showing bilateral variable-sized pulmonary nodules.

6-Pulmonary cysts

Pulmonary cysts **(Figure 6)** are thin-walled air- or fluid-filled spaces. Differentiating pulmonary cysts from centrilobular emphysema on HRCT is important. Emphysematous bullae have no wall and a residual centrilobular artery **[19].** The presence of pulmonary cysts other than honeycomb cysts suggests a diagnosis alternative to idiopathic pulmonary fibrosis (IPF). Pulmonary cysts are often present in pulmonary Langerhans cell histiocytosis, desquamative interstitial pneumonia (DIP), lymphoid interstitial pneumonia, and lymphangioleiomyomatosis **[13-15]**.

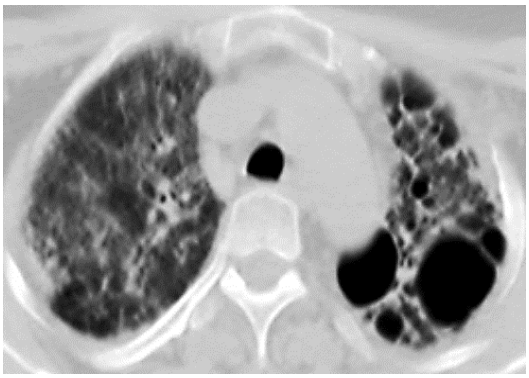

Figure 6: Pulmonary cysts: axial HRCT image showing left-sided thin-walled pulmonary cysts.

7-Ground-glass opacity (GGO)

GGO **(Figure 7)** denotes a homogenous increase of the pulmonary parenchymal attenuation with visible underlying bronchi and vessels. GGO appears less dense than pulmonary consolidations **[13]**. GGO is caused by a thickening of the interstitial septa or partial filling of alveolar spaces with fluids, or cells **[14]**. Minor GGO is considered a portion of the fibrosing process, seen in association with reticulation or traction bronchiectasis **[16]**. The presence of extensive GGO is not a sign of UIP and suggests a diagnosis other than UIP and IPF **[13]**. ILDs presented with predominant GGO include HP, DIP, organizing pneumonia [OP], and alveolar proteinosis **[13-14]**.

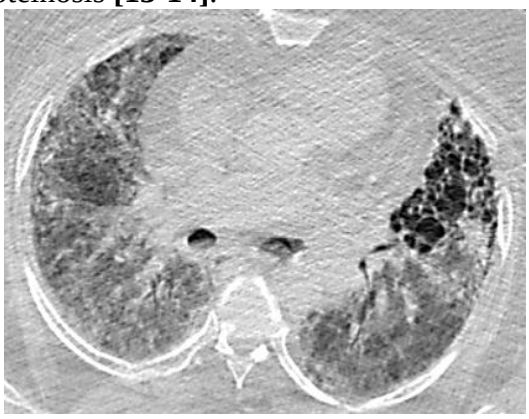

Figure 7: Pulmonary GGO: axial HRCT image showing bilateral basal extensive GGO.

8-Pulmonary consolidation

Pulmonary consolidation **(Figure 8)** indicates a homogenous pulmonary opacity with or without an air bronchogram that obscures the underlying bronchi and vessels. Consolidation is caused by a complete filling of alveolar spaces with fluids, or cells. Consolidation is not a sign of UIP, and predominant consolidation suggests a diagnosis alternative to IPF such as chronic eosinophilic pneumonia and OP **[13-14]**. However, consolidation could be seen in IPF patients complicated with infection or malignancy **[13]**.

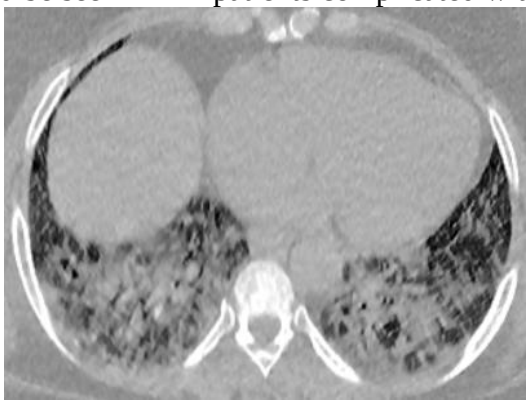

Figure 8: Pulmonary consolidation: axial HRCT image showing bilateral consolidations with an air bronchogram.

9- Mosaic attenuation

Mosaic attenuation **(Figure 9)** describes a heterogeneous attenuation of the pulmonary parenchyma with well-defined borders that correspond to the borders of the secondary pulmonary lobules **[13]**. Mosaic attenuation can be seen in obliterative small airway disease due to air trapping or pulmonary vascular diseases due to different perfusion, or occasionally in ILDs such as HP due to patchy distribution of GGO **[11-20]**.

If there is a decrease in pulmonary vessel caliber in low attenuation pulmonary regions, this refers to the mosaic perfusion pattern. The mosaic perfusion pattern detected in an inspiratory CT scan requires an expiratory scan to confirm or exclude the presence of air trapping **[20]**.

In an expiratory CT scan, if air trapping is present, the low-attenuation pulmonary regions will not change in density, while the high-attenuation regions will increase in density. Conversely, if air trapping is not present, all the pulmonary regions of different attenuations will increase in density at the expiratory CT scan **[11-20]**. Air trapping is most seen in ILD patients diagnosed with HP and sarcoidosis **[13]**.

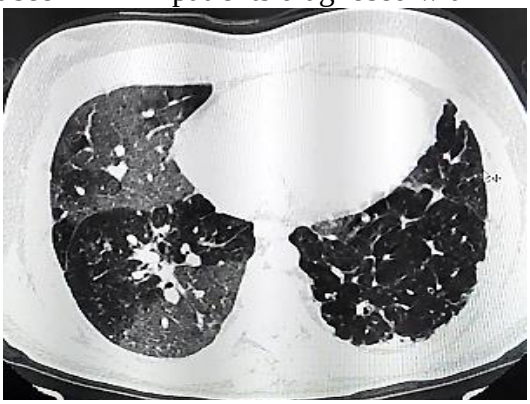

Figure 9: Mosaic attenuation: axial HRCT image showing mosaic attenuation of the right lung.

10-Pulmonary emphysema

Pulmonary emphysema **(Figure 10)** refers to alveolar airspace enlargement and fibrosis-producing thin-walled cystic spaces. Paraseptal emphysema is a common finding in smokers. The differentiation between paraseptal emphysema and honeycombing is sometimes difficult **[16]**. However, emphysematous bullae have thinner walls than honeycomb cysts, seen in a single subpleural layer with a size larger than 1 cm, and are usually seen in association with centrilobular emphysema **[21]**. The presence of combined pulmonary fibrosis and emphysema affects patient management and prognosis, as these patients tend to have worse complications and prognosis. Therefore, the extent and the degree of emphysema should be described in ILD patients at HRCT **[22]**.

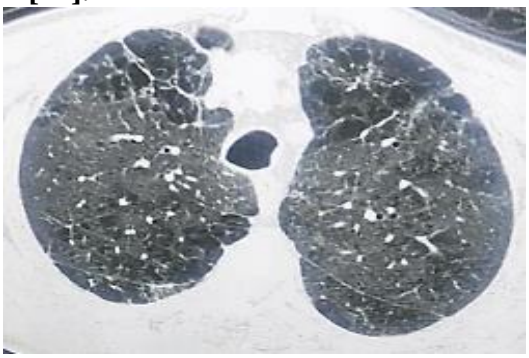

Figure 10: Pulmonary emphysema: axial HRCT image showing hyperinflation of both lungs with pulmonary emphysematous changes, and mosaic appearance.

Distribution of the pulmonary features of ILDs

1- Zonal distribution

Zonal distribution of the pulmonary features of ILDs at HRCT can be determined by dividing the lung into three zones: upper zone, from lung apex to the level of the carina; middle zone, from the carinal level

to the level of inferior pulmonary veins; and lower zone, from inferior pulmonary veins to the level of the diaphragm **[23].**

2- Axial distribution

Axial distribution refers to the distribution of the pulmonary features in the craniocaudal plane from superior to inferior lung regions. Axial distribution of ILD can be described as upper lung–predominant, mid lung–predominant, basal lung predominant, or diffuse axial distribution. Basal and subpleural predominance is most commonly seen in IPF.

3- Apicobasal gradient

The apicobasal gradient **(Figure 11)** indicates an increased intensity of reticulation or honeycombing from the lung apex toward the lung base. It is best seen in coronal reformatted HRCT images and frequently seen in UIP. In NSIP, GGO involves lower lung lobes with no clear apicobasal gradient **[24].**

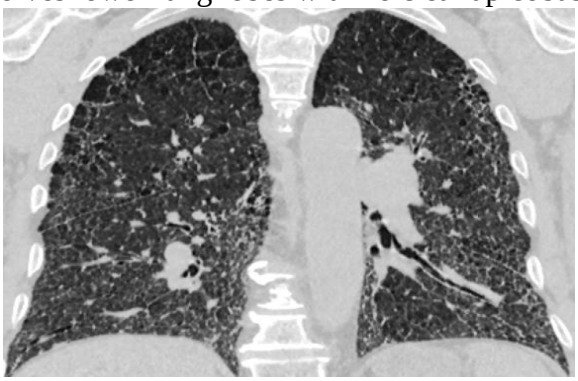

Figure 11: Apicobasal gradient: coronal HRCT image showing an increased intensity of reticulations and honeycombing from the lung apex toward the base.

Pulmonary complications in ILD patients

1- Acute exacerbations of ILDs

Acute exacerbation of ILD describes a clinically significant worsening or decline of respiratory function. It frequently occurs in ILD patients with extensive fibrosis and more in IPF patients than non-IPF patients. It may occur without a clear cause or due to pneumonia, pulmonary embolism, pneumothorax, or cardiac failure **[25].** HRCT imaging findings in acute exacerbation include bilateral diffuse or multiregional GGO with or without diffuse, patchy, or peripheral consolidations. Most ILD patients die within weeks to months after acute exacerbations **[26].**

2- Pulmonary infections

Patients with ILD are at high risk of pulmonary infections, most commonly by tuberculous mycobacteria, aspergillus species with aspergilloma formation in a preexisting cavity. that may show atypical radiologic manifestations as pre-existing pulmonary abnormality may mask the typical radiographic appearances **[18]**.

3-Cancer lung

Prior studies reported a five-fold increase in the incidence of cancer lung in ILD patients compared with the general population especially older male smokers. Adenocarcinoma is the most common histopathological subtype of lung cancer associated with ILD, followed by squamous cell carcinoma. Lung malignancy commonly develops in regions of severe fibrosis at the lung periphery or the junction of the fibrosis and normal lung. Radiologists should carefully evaluate the peripheral fibrotic lung regions for any new or progressive enlarging nodules, lobulated masses, or mass-like consolidations **[26].**

4- Other complications

Spontaneous and usually mild pneumothorax and pneumomediastinum occur in about 12% of patients with ILD. Pulmonary hypertension is common in ILD patients, being identified in about half of ILD patients referred for lung transplantation **[25].**

Categories of ILD-RADS (Table 1)

1-ILD-RADS-0 refers to incomplete or inadequate CT examination

2-ILD-RADS-1 (classic UIP) refers to the patchy distribution of pulmonary reticulations with traction bronchiectasis, honeycombing, and minor GGO with basal and peripheral predominance **[9] (Figure 12).**

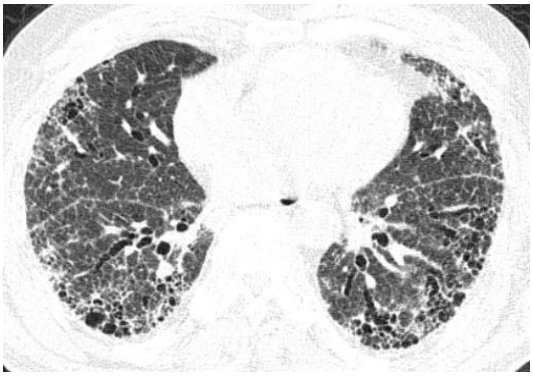

Figure 12: ILD-RADS-1 (typical UIP): HRCT image showing bilateral basal reticulations with traction bronchiectasis and honeycombing.

2- ILD-RADS-2 (possible UIP) refers to the presence of pulmonary reticulations with traction bronchiectasis, and minor GGO in a basal and peripheral predominance without honeycombing **[9] (Figure 13).**

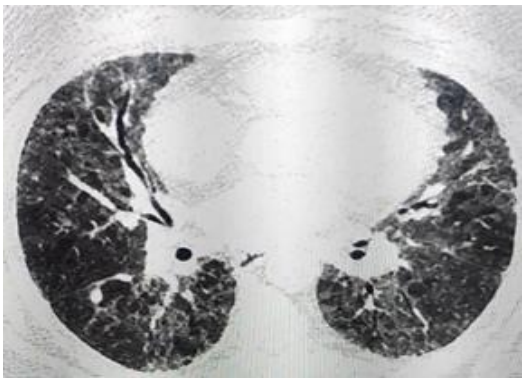

Figure 13: ILD-RADS-2 (possible UIP): HRCT image showing bilateral basal reticulations with traction bronchiectasis with minor GGO and absent honeycombing**.**

3- ILD-RADS-3 (indeterminate for UIP) refers to the diffuse, patchy, or variable distribution of pulmonary reticulations without definite peripheral or basal predominance **[9] (Figure 14).**

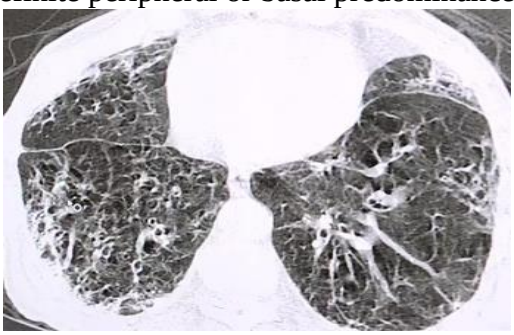

Figure 14: ILD-RADS-3 (indeterminate for UIP): HRCT image showing bilateral reticulations with scattered areas of GGO and absent honeycombing.

4-ILD-RADS 4 (inconsistent with UIP or suggesting an alternative diagnosis) refers to the presence of one or more atypical HRCT features, including cysts, nodules, extensive mosaic attenuation, extensive GGO, or predominant consolidation in a peribronchovascular, perilymphatic axial distribution, or upper or mid-lung zonal distribution **[9] (Figure 15).**

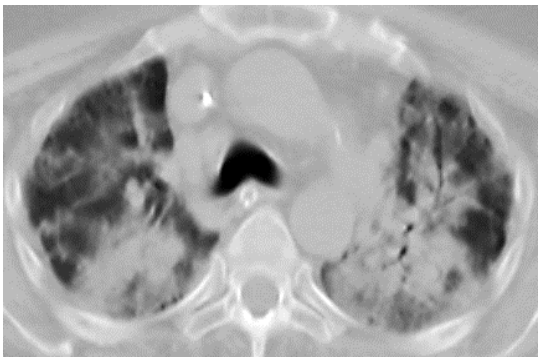

Figure 15: ILD-RADS-4 (suggestive of alternative diagnosis to UIP): HRCT image showing a bilateral upper extensive GGO with consolidation, absent traction bronchiectasis, and honeycombing**.**

Table 1: Categories of ILD-RADS **[9].**

ILD-RADS Category	HRCT features
ILD-RADS-0	No or incomplete CT examination
ILD-RADS-1 (Typical UIP)	Pulmonary reticulations, traction bronchiectasis, honeycombing, and minor ground glass Basal and subpleural predominance No atypical features
ILD-RADS-2 (Probable UIP)	Pulmonary reticulations, traction bronchiectasis, and minor ground glass Basal and subpleural predominance No honeycombing No atypical features
ILD-RADS-3 (Indeterminate for UIP)	Pulmonary reticulations Patchy distribution No basal or subpleural predominance No traction bronchiectasis No honeycombing No atypical features Features and/or distribution do not suggest a specific diagnosis
ILD-RADS-4 (Inconsistent with UIP, suggestive of diagnosis alternative to IPF)	One or more of the atypical features are present: Extensive ground glass Predominant consolidation pulmonary cysts, Extensive mosaic attenuation Diffuse pulmonary nodules or cysts. Upper and middle lung zonal predominance Peribronchovascular axial distribution predominance

Conclusion

HRCT is valuable in the evaluation of ILDs. ILDs have various pulmonary features at HRCT. The correct recognition and interpretation of these features can help in the accurate diagnosis of ILDs. The ILD-RADS provides a standardized template and four categories for the description of ILDs' pulmonary features at HRCT.

References

1. **Wong AW, Ryerson CJ, Guler SA.** Progression of fibrosing interstitial lung disease. Respir Res. 2020;21:32.
2. **Lynch DA, Sverzellati N, Travis WD, et al.** Diagnostic criteria for idiopathic pulmonary fibrosis: a Fleischner society white paper. Lancet Respir Med. 2018;6:138–153.
3. **Park SW, Baek AR, Lee HL, et al.** Korean guidelines for diagnosis and Management of Interstitial Lung Diseases: part 1. Introduction Tuberc Respir Dis. 2019;82:269–276.
4. **Cosgrove GP, Bianchi P, Danese S, Lederer DJ**. Barriers to timely diagnosis of interstitial lung disease in the real world: the INTENSITY survey. BMC Pulm Med 2018;18:9.
5. **Collins BF, Luppi F.** Diagnosis and management of fibrotic interstitial lung diseases. Clinics in Chest Medicine. 2021 Jun 1;42(2):321-35.

6. **Cottin V, Hirani NA, Hotchkin DL, et al.** Presentation, diagnosis and clinical course of the spectrum of progressive-fibrosing interstitial lung diseases. Eur Respir Rev.2018;27(150).
7. **Rabahi MF, Moreira MA, Escuissato DL, Meirelles GD, Marchiori E**. Importance of chest HRCT in the diagnostic evaluation of fibrosing interstitial lung diseases. Jornal Brasileiro de Pneumologia 2021;31:47.
8. **Dsouza K. and de Andrade, JA.** The Diagnostic Approach to Interstitial Lung Disease. Current Pulmonology Reports, **2018;** 7:149.
9. **Berkowitz EA, Bernheim A, Little BP**. Introducing ILD-RADS: a pilot study of an interstitial lung disease standardized reporting template. J Am Coll Radiol. 2019;16:1169–1172.
10. **Gruden JF, Naidich DP, Machnicki SC, et al**. An algorithmic approach to the interpretation of diffuse lung disease on chest CT imaging: a theory of almost everything. Chest. 2020;157:612–635.
11. **Chung JH, Landeras L**. Probable UIP: What is the evidence that compels this classification and how is it different from the indeterminate category? Semin Roentgenol. 2019;54:15–20.
12. **Chen L, Halai V, Leandru A, et al.** Interstitial lung disease: update on the role of computed tomography in the diagnosis of idiopathic pulmonary fibrosis. J Comput Assist Tomogr. 2019;43:898–905.
13. **Hobbs S, Chung JH, Leb J, Kaproth-Joslin K, Lynch DA**. Practical imaging interpretation in patients suspected of having idiopathic pulmonary fibrosis: official recommendations from the Radiology Working Group of the Pulmonary Fibrosis Foundation. Radiology: Cardiothoracic Imaging. 2021;3(1):e200279.
14. **Hochhegger B, Marchiori E, Zanon M, et al**. Imaging in idiopathic pulmonary fibrosis: diagnosis and mimics. Clinics. 2019;74:e225.
15. **Jones KD.** Pulmonary cystic disease and its mimics. Surg Pathol Clin.2020;13:141–163.
16. **MartinMD, Chung JH, Kanne JP.** Idiopathic pulmonary fibrosis. J Thorac Imaging. 2016;31:127–139.
17. **Glass LN, Sumon M, Goulart H, et al.** Disappearing nodules: spontaneously regressing pulmonary amyloidosis. BMJ Case Rep.2019;12.
18. **Chiarenza A, Esposto Ultimo L, Falsaperla D, et al.** Chest imaging using signs, symbols, and naturalistic images: a practical guide for radiologist and non-radiologists. Insights Imaging. 2019;10:114.
19. **Lee KC, Kang EY, Yong HS, et al.** A stepwise diagnostic approach to cystic lung diseases for radiologists. Korean J Radiol. 2019;20:13681380.
20. **Palleiro AG, Mazzini SP, Franquet T**. Basic HRCT patterns in diffuse interstitial lung disease. Radiología (English Edition). 2022;64:215-26.
21. **Digumarthy S, Abbara S, Chung J**. Problem solving in chest imaging. Philadelphia, Pa: Elsevier, 2020.
22. **Salvatore M, Smith ML.** Cross sectional imaging of pulmonary fibrosis translating pathology into radiology. Clin Imaging. 2018;51:332–336.
23. **Key AL, Holt K, Warburton CJ, et al.** Use of zonal distribution of lung crackles during inspiration and expiration to assess disease severity in idiopathic pulmonary fibrosis. Postgrad Med J. 2018;94:381–385.
24. **Hatabu H, Hunninghake GM, Lynch DA.** Interstitial lung abnormality: recognition and perspectives. Radiology. 2019;291:1–3.
25. **Baratella E, Fiorese I, Marrocchio C, et al.** Imaging Review of the Lung Parenchymal Complications in Patients with IPF. Medicina. 2019;55.
26. **Fukui M, TakamochiK, SuzukiK, et al.** Lobe specific outcomes of surgery for lung cancer patients with idiopathic interstitial pneumonias. Gen Thorac Cardiovasc Surg. 2020.