HRCT imaging findings of diffuse lung involvement in collagen vascular diseases

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Learning objectives

• To briefly review the most common collagen vascular diseases (CVDs) which may have pulmonary involvement.
• To recognize typical and atypical chest High-Resolution Computed Tomography (HRCT) pulmonary and extrapulmonary imaging findings.
Background

CVDs constitute a group of autoimmune disorders, whose common denominator is the damage to components of connective tissue, frequently targeting pulmonary interstitium, airways, pleura, and pulmonary vasculature.\(^\text{(1,2)}\) Diagnosis of these conditions is often challenging, while management concentrates on controlling inflammation and preventing damage, since most of them respond well to immunosuppressant therapy.\(^\text{(3,4,5)}\)

The two thoracic manifestations with the greatest clinical impact in patients with CVDs are interstitial lung disease (ILD) and pulmonary artery hypertension (PAH).\(^\text{(4,6)}\) CVDs most commonly associated with ILD include Systemic Lupus Erythematosus (SLE), Rheumatoid Arthritis (RA), Progressive Systemic Sclerosis/Scleroderma (SSc), Sjögren Syndrome (SS), Dermatomyositis/Polymyositis (DM/PM) and Mixed Connective Tissue Disease (MCTD).\(^\text{(7)}\) Conversely, about 15% of patients presenting for evaluation of ILD have an underlying CVD, since respiratory symptoms can be the first clinical manifestation.\(^\text{(4,8)}\)

The most common patterns of ILD encountered in patients with CVD include: usual interstitial pneumonia (UIP), non-specific interstitial pneumonia (NSIP), organizing pneumonia (OP), lymphoid interstitial pneumonia (LIP), and acute interstitial pneumonia (AIP).\(^\text{(9)}\) Table 1 (Table 1 on page 4) resumes usual and unusual HRCT related findings, along with their typical distribution. Medications used to treat CVDs may result in collateral lung injury, which must be distinguished from CVD-related ILD; furthermore, immunosuppressive drugs can predispose patients to opportunistic pulmonary infections, which can confound pulmonary findings.\(^\text{(4,5,10)}\)

Pulmonary involvement is an important prognostic factor in CVDs, especially if PAH is associated.\(^\text{(10,11)}\) Therefore, early identification of CVD-related ILD is essential in order to guide prompt patient management. HRCT is a key diagnostic tool to detect, characterize and quantify the extent of ILD. Imaging can also identify extrapulmonary sites of CVD involvement.\(^\text{(2,10-12)}\)
Table 1: Usual and unusual HRCT findings in CVDs, along with their typical distribution

<table>
<thead>
<tr>
<th>HRCT Pattern</th>
<th>Histopathologic Findings</th>
<th>Radiologic Findings</th>
<th>Typical findings</th>
<th>Typical distribution</th>
<th>Unusual findings</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Usual interstitial Pneumonia (UIP)</strong></td>
<td>Architectural destruction, fibrosis with honeycombing, fibroblastic foci; alternating areas of normal lung tissue, inflammation, fibrosis, and honeycombing (temporally heterogeneous)</td>
<td></td>
<td>Reticular opacities, honeycombing, traction bronchiectasis or broncholectasis, architectural distortion, focal ground glass attenuation</td>
<td>Peripheral, subpleural, basal</td>
<td>Asymmetrical distribution, major involvement of upper lobes</td>
</tr>
<tr>
<td><strong>Non-specific interstitial Pneumonia (NSIP)</strong></td>
<td>Varying proportion of interstitial inflammation and fibrosis, divided into cellular (inflammation) and fibrosing (fibrosis) patterns; patchy with intervening normal lung tissue; temporally uniform</td>
<td></td>
<td>Ground-glass attenuation, irregular lines, traction bronchiectasis, consolidation</td>
<td>Peripheral, basal, symmetric</td>
<td>Extensive reticular pattern, consolidation (may represent areas of co-existing organizing pneumonia), peribronchovascular distribution</td>
</tr>
<tr>
<td><strong>Organizing pneumonia (OP)</strong></td>
<td>Intraluminal organizing fibrosis in distal airspaces, patchy, preservation of lung architecture, temporally uniform, mild chronic interstitial inflammation</td>
<td></td>
<td>Patchy consolidation or nodules, peri-lobular pattern, reverse halo sign</td>
<td>Subpleural or peribronchial</td>
<td>Focal consolidation mimicking mass may progress to fibrosis</td>
</tr>
<tr>
<td><strong>Lymphocytic interstitial Pneumonia (LIP)</strong></td>
<td>Infiltration of T lymphocytes, plasma cells, and macrophages; lymphoid hyperplasia; diffuse; predominantly septal</td>
<td></td>
<td>Centrilobular nodules, ground-glass attenuation, septal and bronchovascular thickening, thin-walled cysts</td>
<td>More commonly, lower lung predominant</td>
<td>Isolated pulmonary cysts, ground-glass opacity as an isolated finding, septal thickening, and centrilobular nodules</td>
</tr>
<tr>
<td><strong>Acute interstitial pneumonia (AIP)</strong></td>
<td>Alveolar edema, hyaline membrane, fibroblastic proliferation with little mature collagen, diffuse, temporally uniform</td>
<td></td>
<td>Consolidation and ground-glass opacity, often with lobular sparing; traction bronchiectasis later</td>
<td>Diffuse or patchy</td>
<td>May progress to pulmonary fibrosis. Patients who survive may have HRCT pattern similar to non-specific interstitial pneumonia</td>
</tr>
</tbody>
</table>

Table 1: Usual and unusual HRCT findings in CVDs, along with their typical distribution.

Findings and procedure details

1. Indications to HRCT

HRCT is considered the imaging technique of choice for the detection and characterization of ILD in patients with CVD. Indications to HRCT in patients with CVD include:

• baseline evaluations at CVD diagnosis when pulmonary involvement is suspected;
• monitoring of therapy when pulmonary involvement is proved;
• targeting of biopsy.\(^{(7,13,14)}\)

2. HRCT Technique

The use of thin-section CT images (slice thickness ranging from 0.625 mm to 1.5 mm) with a high spatial frequency reconstruction algorithm (e.g., bone or lung algorithms) defines HRCT. The key technical requirement for ‘best practice’ is helical scan, since volumetric data sets give contiguous thin section images of the entire chest with essentially isotropic voxels, thus enabling: a) multiplanar reformatting (without loss of spatial resolution), which facilitates evaluation of the distribution of diffuse lung disease (Fig. 1 on page 12 and Fig. 2 on page 12); b) Maximum Intensity Projection (MIP) and Minimum Intensity Projection (MinIP) reconstructions, which improve the identification of ancillary findings (e.g., lung nodules), the characterization of patchy ILD and the evaluation of their distribution and extent. Importantly, volumetric HRCT scan allows for the crucial differentiation between honeycombing and traction bronchiectasis.\(^{(15-17)}\)

Image data are routinely acquired at suspended full inspiration in the supine position. Additional options include obtaining inspiratory prone and/or end-expiratory scans. The frequent finding of an amorphous increase in attenuation of the dependent lung (i.e., the postero-basal segments of the lower lobes in the supine position) may mimic subtle interstitial abnormalities; prone imaging may help in differentiating posterior lung disease from dependent atelectasis in such cases (Fig. 3 on page 13). In patients with SSc, some authors suggest performing a stand-alone HRCT scan in prone position, or a low sampling protocol obtained in prone position at end inspiration for screening ILD.\(^{(18)}\) In patients with obstructive pulmonary disease, dynamic HRCT during forced expiration demonstrates the presence of regions with inhomogeneous increase in lung attenuation, indicative of air trapping (Fig. 4 on page 14).\(^{(19-20)}\)

The HRCT protocol in use at our Institution is illustrated in Table 2 (Table 2 on page 26).
**Low-dose scanning** - Since the major drawback of the volumetric HRCT technique is the high radiation dose exposure to counteract the noise inherent in thin sections, lower dose techniques have been investigated. Such modified protocols (e.g., 40-80 mAs) demonstrated good diagnostic accuracy in patients with chronic diffuse infiltrative lung disease, even if a potential underestimation of subtle ground glass opacities and emphysema must be taken into account.\(^{(21,22)}\) Low-dose techniques are particularly useful when additional prone and/or expiratory scans are required, and when examining young patients, especially if frequent follow-up is planned.\(^{(16,17,19)}\)

**MIP** - The primary clinical application of MIP is to improve the detection of small pulmonary nodules, their profusion and their distribution (i.e., perivascular vs. centrilobular) \(\text{(Fig. 5 on page 15)}\). In addition, MIP sections of variable thickness are excellent for assessing size and location of pulmonary arteries and veins: pulmonary oedema associated with enlarged pulmonary veins can easily be differentiated from other causes of diffuse ground glass attenuation.\(^{(15,16,23)}\)

**MinIP** - It is the optimal tool for better detect, localize and quantify air trapping \(\text{(Fig. 4 on page 14)}\) and honeycombing \(\text{(Fig. 1 on page 12, Fig. 6 on page 16 and Fig. 7 on page 17)}\), better displaying the subtle difference in density between air and lung parenchyma. MinIP can also be useful in evaluating distribution of cysts \(\text{(Fig. 8 on page 18)}\) and bronchiectasis \(\text{(Fig. 9 on page 19)}\).\(^{(15,16,23)}\)

### 3. Imaging Patterns and differential diagnosis

Since in most cases HRCT appearance is nonspecific, radiologic findings should never be interpreted without knowledge of the clinical picture. Nevertheless, in certain clinical circumstances, HRCT findings can suggest the diagnosis of a specific CVD. The best approach to evaluate CVD is to recognize and analyze different patterns of involvement, which include pulmonary, pleural and mediastinal lymph nodes involvement.

The main HRCT finding in patients with CVDs is diffuse lung involvement. Different HRCT abnormalities reflect the relative proportions of lung fibrosis and inflammation: a reticular pattern with traction bronchiectasis mirrors a predominantly fibrotic process, whereas a predominant ground glass attenuation pattern corresponds to an inflammatory process.\(^{(1)}\)

Other radiologic signs suggestive of underlying CVD include oesophageal abnormalities, pleural and pericardial effusions and thickening, pulmonary arterial enlargement, mediastinal lymphadenopathy, and osteoarticular disease (e.g., glenohumeral joints' involvement in RA).\(^{(1-4,7,10)}\) Table 3 \(\text{(Table 3 on page 27)}\) resumes the frequency of the various ILD patterns according to specific CVDs.
**Rheumatoid arthritis (RA)**- RA can affect many organs, and predominantly involves the synovial tissues and joints. It occurs more frequently in women (female:male ratio of 3:1), typically between 25 and 50 years of age.\(^{(2)}\)

Pleuro-pulmonary complications are common and include interstitial pneumonitis and fibrosis, rheumatoid nodules, bronchiectasis, bronchiolitis and pleural effusion or thickening.\(^{(2,4,7,24,25)}\)

The predominant ILD pattern is UIP (in up to 56% of patients, *Fig. 1 on page 12*) followed by NSIP and OP. Primary utility of HRCT is the reliable identification of patients with UIP-type ILD, who carry a worse prognosis that is similar to IPF. Conversely, the airway-predominant pattern manifests as bronchial wall thickening, bronchiectasis and mosaic attenuation with air trapping.\(^{(4)}\)

Pulmonary macronodules (*Fig. 10 on page 20*), typically asymptomatic, are commonly present in the upper and mid peripheral lung zones. Nodules may cavitate, enlarge, stay stable, spontaneously resolve, or newly appear on serial imaging; due to their nonspecific appearance, follow-up imaging, bronchoscopy or even biopsy are needed.\(^{(2)}\)

**Progressive Systemic Sclerosis/Scleroderma (SSc)** - SSc is characterised by widespread inflammation and fibrosis that produces scar tissue in the skin, internal organs and small blood vessels. Age of onset is most commonly in the range of 30-50 years, with a female:male ratio of 5-14:1.\(^{(3)}\)

On HRCT, pulmonary manifestations consisting of ILD can be found in 55-65% of patients. The most common pulmonary pattern is NSIP (*Fig. 4 on page 14*), whereas UIP is less frequently observed (*Fig. 6 on page 16*). Findings at initial presentation carry prognostic significance: patients with extensive ILD, occupying 20% or more of lung volume, experience a rapid decline in pulmonary function, whereas patients with less extensive lung abnormality or no pulmonary involvement have relatively little progression of ILD at follow-up.\(^{(1,2,4,7,24,25)}\)

Several HRCT extrapulmonary thoracic findings help suggest the diagnosis of SSc: oesophageal involvement, presenting as a dilated patulous oesophagus often containing fluid or debris, is reported in up to 97% of patients (*Fig. 11 on page 21*). Imaging may also demonstrate stigmata of aspiration because of oesophageal dysmotility, including bronchiectases (*Fig. 3 on page 13*), mucus plugging and tree-in-bud nodularities.\(^{(2)}\)

Enlargement of central pulmonary arteries suggesting PAH may also be seen (*Fig. 12 on page 22*), as a result of restrictive lung disease or CVD-related vasculopathy in the absence of ILD. The ratio between the diameters of cardiac ventricles may also have prognostic values in these patients.\(^{(2)}\)
Sjögren syndrome (SS)- SS is a slowly progressive, inflammatory disease affecting primarily the exocrine glands. SS affects mostly females (female:male ratio 9:1) during the fourth and fifth decades of life.(3)

Thoracic involvement is common, including airway disease and ILD. Since patients with primary SS have an increased rate of respiratory infections, differentiate scarring sequel from parenchymal abnormalities related to ILD can be difficult on imaging.

The most common HRCT pattern in patients with SS-related ILD is NSIP (Fig. 2 on page 12), while OP (Fig. 13 on page 23), LIP and UIP (Fig. 7 on page 17) are less frequent.(1-4,7,24,25) Thin-walled parenchymal cysts and small peripheral nodules can raise suspicion for LIP, particularly when ground glass opacities are present. Cystic lung disease associated with pulmonary amyloidosis has also been described (Fig. 8 on page 18).(26-28)

Airway involvement is common in patients with SS, with bronchiectasis (Fig. 9 on page 19), mucus plugging and tree-in-bud nodules (Fig. 5 on page 15), potentially resulting in an obstructive pattern at spirometry.(1-4,7,24,25)

SS is associated with an increased risk of malignant non-Hodgkin's lymphoma, frequently involving extranodal sites, most commonly the parotid glands but also the lung. The HRCT differential diagnosis between lymphoma and LIP is challenging, stating a similar prevalence of findings such as lung masses, nodules, ground glass opacities, cavities, pericardial effusion, and hilar lymphadenopathies.(2)

An association between SS and thymoma also been reported (Fig. 14 on page 24).(29,30)

Systemic Lupus Erythematosus (SLE) - SLE is characterised by widespread inflammation. Peak incidence occurs between 15 and 40 years of age, with a female:male ratio of 6-10:1. Pleuro-pulmonary involvement occurs in approximately 50-60% of patients.(3)

HRCT typically demonstrates lower lobe-predominant septal thickening, architectural distortion, and ground glass opacities suggestive of NSIP pattern, with or without pleural effusion. In the acute setting, HRCT findings can range from minimal abnormality to diffuse ground glass opacities, patchy consolidation or both, suggesting diffuse alveolar damage (DAD). Diffuse alveolar haemorrhage is another acute finding, manifesting as nonspecific diffuse alveolar opacities and often accompanied by haemoptysis. Airway (i.e., bronchiectasis) and vascular involvement (including stigmata of PAH) are frequently associated findings, reported in up to 20% and in up to 11% of
The occurrence of bronchus-associated lymphoid tissue lymphoma (BALT) has also been observed (Fig. 15 on page 25).

Patients with SLE may also frequently present with pleuritic chest pain; thoracentesis with pleural fluid analysis, including the presence of antinuclear antibodies, proved to be helpful in diagnosing lupus pleuritis. Other manifestations of lupus serositis include pericarditis, occasionally presenting with significant pericardial effusion.

**Dermatomyositis/Polymyositis (DM/PM)** - DM/PM are systemic inflammatory disorders with a bimodal distribution, with peaks between 10-15 and 45-60 years of age (malignancy associated to myositis and inclusion body myositis is more common in the second peak). The female: male ratio is about 2.5:1.

The rate of ILD is variable, ranging from 5-30% to as high as 65% or more in HRCT studies, with many asymptomatic patients having imaging findings of ILD. HRCT findings generally depict an NSIP or OP pattern. Patients with PM-DM may also develop respiratory complications, such as hypoventilation from respiratory muscle weakness and aspiration from pharyngeal muscle weakness.

Relatively few extrapulmonary thoracic manifestations of PM-DM have been described.

**Overlap Syndrome/Mixed Connective Tissue Disease (MCTD)** - Overlap syndrome is a disorder in which features of various CVDs can coexist and overlap.

Pulmonary abnormalities on imaging are a common feature of MCTD (52-85% of patients) and ILD is the most common manifestation (mainly NSIP), varying from asymptomatic involvement to advanced fibrosis. Other pulmonary findings may include low lung volumes related to respiratory muscle weakness, stigmata of aspiration or pneumonia or both, and rarely alveolar haemorrhage.

Extrapulmonary thoracic findings include signs of PAH (in 10-45% of patients), signs of oesophageal dysmotility (as with SSc) and pleural and pericardial effusions or thickening.

**HRCT ILD-Patterns: tips for differential diagnosis**

The different HRCT patterns of CVD-related ILDs mirror those seen in the corresponding idiopathic interstitial pneumonias, with some subtle peculiarities in presentation.
Findings typical of **UIP** pattern include reticulation (which predominates in peripheral and basilar regions), honeycombing and traction bronchiectasis, often with spatial and temporal heterogeneity. UIP in the setting of CVD tends to have less honeycombing than idiopathic UIP, fewer fibroblast foci on histologic analysis and a somewhat better prognosis.\(^{(4,33)}\)

HRCT findings in **NSIP** differ from UIP in that ground glass (often with subpleural sparing) predominates over honeycombing. Although subpleural sparing may be helpful in diagnosing NSIP, it is not a requirement. NSIP tends to be more spatially and temporally homogeneous than UIP (Fig. 2 on page 12 and Fig. 6 on page 16).\(^{(4,34)}\)

**OP** typically demonstrates patchy peripheral airspace opacities in the subpleural or peribronchovascular regions that vary in attenuation from ground glass to dense consolidation and may be band-like, polygonal (Fig. 13 on page 23), or nodular in shape. Crescentic opacities surrounding a focus of ground glass, known as the "atoll" or "reverse halo sign", may be helpful in suggesting the diagnosis. The presence of OP superimposed on other patterns of ILD, especially NSIP, is very suggestive of an underlying CVD.\(^{(4,26-28)}\)

**LIP** is classically associated with SS and typically manifests as ground glass with poorly defined centrilobular nodules and scattered thin-walled cysts on HRCT images. Cysts associated with calcified nodules may represent amyloid deposits. Possible coexistence of LIP in patients with amyloid-associated cystic lung disease cannot be excluded, especially if histopathologic specimens are limited (Fig. 8 on page 18).\(^{(4,26-30)}\)

**AIP** commonly consists in an air space disease (consolidation and ground glass) that is usually a manifestation of underlying DAD. This pattern usually occurs in the setting of an acute exacerbation of ILD, potentially complicating nearly every form of CVD-related ILD. When an acute exacerbation manifests as DAD, the prognosis is typically poor. AIP usually superimposes on an underlying pattern of ILD, such as UIP or NSIP, but can occasionally be the initial pulmonary manifestation of CVD.\(^{(4,36,37)}\)

**4. Impact of HRCT on clinical management**

The two thoracic manifestations with the greatest clinical importance in patients with CVDs are interstitial lung disease (ILD) and pulmonary artery hypertension (PAH), which are responsible for a large part of the mortality and morbidity in this patients group.

Before offering a diagnosis of thoracic involvement in CVD, the radiologist must exclude the possibilities of drug-related complications and opportunistic infections. Correlation of HRCT findings with clinical and pathological features is required, since most drug-induced immune reactions are diagnosed *per exclusionem*.
ILD is a challenging clinical entity that can be associated with various CVDs. Timely imaging and accurate detection of pulmonary involvement in CVDs have important therapeutic and prognostic implications, since prompt treatment leads to improved outcome. A collaborative and multidisciplinary approach to CVDs management is essential. Serial imaging, in conjunction with pulmonary function tests, can be used to follow severity of pulmonary involvement and to guide management. Precise knowledge of HRCT signs of thoracic involvement in CVDs allows radiologists to provide better guidance for treatment and follow-up.\textsuperscript{(6, 8, 11-14, 38)}
Fig. 1: UIP-pattern in a 78-year-old female patient with Rheumatoid Arthritis. HRCT image on the axial plane (A) shows subpleural, irregular, linear hyperattenuating areas, more conspicuous in the posterior aspect of the right inferior lobe, suggesting initial honeycombing. HRCT image reformatted on the coronal plane (B) confirms initial honeycombing. HRCT 8 mm thick Minimum Intensity Projection (MinIP) slab image on the coronal plane (C) better depicts and quantifies honeycombing.

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Fig. 2: NSIP-pattern in a 75-year-old female patient with Sjögren Syndrome. HRCT image on the axial plane (A) shows bilateral symmetrical ground glass opacities in a basal and peripheral distribution with associated hyperattenuating reticulation. HRCT image reformatted on the coronal plane (B) better depicts the spatial distribution of such findings.

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**Fig. 3:** Initial NSIP-pattern in a 57-year-old female patient with Progressive Systemic Sclerosis/Scleroderma. HRCT image on the axial plane in supine position (A) depicts subtle interstitial abnormalities in the posterior aspects of lower lobes, which are confirmed in prone position (B).

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**Fig. 4:** Air trapping in a 66-year-old female patient with Progressive Systemic Sclerosis/Scleroderma. (A) Inspiratory HRCT image on the axial plane barely shows a pattern of subtle mosaic attenuation in both lower lobes. (B) End-expiratory scan at the same level clearly enhances air trapping. HRCT 10 mm thick Minimum Intensity Projection (MinIP) slab images on the coronal plane (C at suspended full inspiration, and D at end-expiratory) helps to detect, localize and quantify mosaic attenuation.

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Fig. 5: Bronchiectasis and mucoid impaction with concomitant tree-in-bud opacities in a 77-year-old female patient with Sjögren Syndrome. HRCT image on the axial plane (A) shows mild cylindrical dilatation of peripheral bronchi in the right upper lobe. HRCT 8 mm thick Maximum Intensity Projection (MIP) slab image on the axial plane (B) improves evaluation of tree-in-bud opacities distribution.

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**Fig. 6:** UIP-pattern in a 73-year-old male patient with Progressive Systemic Sclerosis/Scleroderma. HRCT image on the axial plane (A) shows asymmetrical basal and peripheral honeycombing with associated mild ground glass opacities, traction bronchiectasis and architectural distortion. HRCT 10 mm thick Minimum Intensity Projection (MinIP) slab images on the coronal (B) and sagittal (C) planes well depicts honeycombing distribution in lower lobes and left upper lobe. Patulous oesophagus containing fluid is also evident.

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Fig. 7: UIP-pattern in a 57-year-old male patient with Sjögren Syndrome. HRCT image on the axial plane (A) shows basal and peripheral reticulation with associated ground glass opacities, mild honeycombing, traction bronchiectasis and architectural distortion, reflecting lung fibrosis. HRCT 10 mm thick Minimum Intensity Projection (MinIP) slab images on the coronal (B) and sagittal (C) planes well depict honeycombing distribution in lower lobes.

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**Fig. 8:** LIP-pattern in a 77-year-old female patient with Sjögren Syndrome, complicated by amyloidosis and thymoma. HRCT images on the axial plane (A,B) shows multiple thin-walled cysts in both lungs (arrowheads in A) and irregularly multiple nodules within the middle right lobe (arrows in A,B). Biopsy of one of these nodules revealed amyloid deposits at pathology. HRCT 8 mm thick Minimum Intensity Projection (MinIP) slab image on the coronal plane (C) better evaluates distribution of cysts.

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Fig. 9: Bronchiectasis in a 53-year-old female patient with Sjögren Syndrome. HRCT images on the axial plane (A,B) show mild cylindrical dilatation of peripheral bronchi in both lower lobes (arrows in A,B). HRCT image reformatted on the coronal plane (C) confirms bibasal distribution of these findings. HRCT 20 mm thick Minimum Intensity Projection (MinIP) slab image on the coronal plane well depicts bronchiectasis in lower lobes, associated with irregular linear hyperattenuating disventilatory areas.

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Fig. 10: Multiple rheumatoid nodules in two different female patients with Rheumatoid Arthritis. (A-D) HRCT images on the axial plane show multiple bilateral macronodules, some with internal cavitation (arrows in B-D). Biopsy demonstrated granulomatous changes suggestive of a rheumatoid nodule at pathology.

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Fig. 11: Patulous oesophagus (red star) in a 59-year-old female patient with Progressive Systemic Sclerosis/Scleroderma and lung abnormalities suggestive of a NSIP-pattern.

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Fig. 12: Pulmonary hypertension in the same patient of Figure 6. CT image on the axial plane demonstrates an enlarged pulmonary artery (34 mm diameter, red star).

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**Fig. 13:** OP-pattern in a 66-year-old female patient with Sjögren Syndrome. HRCT image on the axial plane shows bilateral poligonal consolidation areas in the upper lobes (arrows), with peripheral distribution and with associated air bronchograms and adjacent ground glass opacities. The lesions disappeared after corticosteroid treatment.

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Fig. 14: Thymoma in the same patient of Figure 8. Post-contrast CT image on the axial plane shows a large anterior mediastinal mass (red star), with smooth margins and homogeneous attenuation.

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**Fig. 15:** Biopsy-proven marginal zone B-cell lymphoma of BALT in a 71-year-old female patient with Systemic Lupus Erythematosus. HRCT image on the axial plane shows a patchy area of consolidation in the right upper lobe, containing air bronchograms.

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<table>
<thead>
<tr>
<th>Scan type</th>
<th>Helical full</th>
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<tr>
<td>kVp</td>
<td>100-120</td>
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</table>
| mA                | Suspended full inspiration: 150-400 (Automatic Modulation)  
|                   | Additional acquisition: 150-400 (Automatic Modulation) |
| Rotation time (s) | 0.8         |
| Beam width (mm)   | 40          |
| Transaxial reconstruction slice thickness (mm) | 1.25 |
| Field of view     | Both lungs, from base to apex |
| Noise index       | 18.4        |
| Acquisition       | Suspended full inspiration in the supine position.  
|                   | Additional scans in prone position or during forced expiration |
| WW (H.U.)         | 1700 (lung), 350 (soft tissues) |
| WL (H.U.)         | -550 (lung), 50 (soft tissues) |
| Reconstruction algorithm | HD bone plus |

**Table 2:** Technique for Volumetric HRCT of Diffuse Lung Diseases

Table 2: HRCT protocol in our Institution, with a 64-slice multidetector CT scanner (LightSpeed HD Discovery 750, General Electrics, Milwaukee, USA).

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<th></th>
<th>RA</th>
<th>SSc</th>
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**Table 3**: Frequency of the various ILD patterns according to specific CVDs.

Conclusion

- Chest HRCT allows assessment of interstitial lung involvement and of extrapulmonary manifestations in patients with CVDs; most cases show a non-specific interstitial pneumonia (NSIP) pattern.
- Chest HRCT features of CVDs may suggest clinical entities and influence management before final diagnosis.


