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Multidisciplinary working group for interstitial lung disease in Thailand: Part 3 – the proposed visual scoring method for quantifying the global disease and fibrotic extents on high-resolution CT

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Introduction

Interstitial lung disease (ILD) encompasses various pulmonary parenchymal disorders [1]. Some ILDs, especially idiopathic interstitial fibrosis (IPF) and systemic sclerosis-associated ILD, can become progressive and devastating [1]. Progressive fibrosing ILD is suggested when patients having any of the following criteria have experienced disease progression in 24 months. These criteria [1] include

1. A relative decline of ≥10% in forced vital capacity (FVC);
2. A relative decline of ≥15% in diffusing capacity of the lung for carbon monoxide (*D*LCO); or
3. Worsening symptoms or a worsening radiological appearance accompanied by a ≥5–<10% relative decrease in FVC.

Early detection of ILD provides the opportunity for early therapeutic intervention, which could improve patient outcomes [1-3]. It is now generally accepted that visual abnormalities correlate with the extent of pathological involvement and the severity of physiologic impairment. Hence, determining the extent of visual abnormalities is increasingly essential for managing ILD patients, monitoring disease progression, response to therapy, and eligibility for novel antifibrotic therapies [1-3].

The ILD extent on high-resolution computed tomography (HRCT) can be quantified by visual scoring methods or quantitative CT analysis tools [4, 5]. However, due to limited resources for quantitative CT analysis, visual scoring methods for quantifying the ILD severity and the extent from HRCT remain essential.

Our previous article [6] summarizes various visual scoring methods published in the literature. Since some anatomical levels described in the previously published methods can be subject to individual interpretations [4, 6-8], we proposed a new visual scoring method for quantifying the global disease extent and the fibrotic extent on HRCT made by the consensus of the multidisciplinary working group for ILD in Thailand [9].

Proposed visual scoring method for quantifying the ILD extent on HRCT

The proposed visual scoring method estimates the ILD extent on HRCT in terms of the global disease extent and the fibrotic extent. Evaluation of the global disease extent helps evaluate responses (reduction or resolution of active inflammation) following immunosuppressive treatment.

The global disease extent denotes the extent of all parenchymal abnormalities related to ILD, including lung fibrosis (reticular opacities, traction bronchiolectasis, traction bronchiectasis, honeycombing, and architectural distortion) and areas with active inflammation or acute exacerbation seen as pure ground-glass opacity or consolidation [10]. The global disease extent of the ILD should not include unrelated abnormalities, e.g., active infection, concomitant emphysema secondary to cigarette smoking, pulmonary edema, mosaic perfusion, dependent atelectasis, or pseudo-ground-glass opacities caused by expiration [10-12].

Unlike the global disease extent, the fibrotic extent denotes the extent of the abnormalities showing irreversible parenchymal scarring or fibrotic changes associated with ILD. Therefore, the fibrotic extent comprises the lung areas showing reticular opacities (thickened intralobular or interlobular septa), traction bronchiolectasis, traction bronchiectasis, honeycombing, and architectural distortion [4, 10, 13, 14]. The fibrotic extent should not include any fibrotic scarring or cicatricial atelectasis due to other unrelated conditions, such as remote infection, trauma, or surgery.

Dissimilar to the previously published visual scoring methods [6], this proposed method estimates the percentage of lung involvement at the six predefined anatomical lung levels shown on HRCT. The predefined anatomical lung levels 1-5 are adopted from the previously published methods [7, 8]. Due to basal-predominant ILD, level 6 is intentionally added for a more accurate estimation of the disease extent. For a precise depiction of the predefined anatomical levels, volumetric HRCT scanning is preferred [10].

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The six predefined levels (Figure 1) are as follows: 1) the origin of the great vessels, immediately above the top of the aortic arch; 2) the main carina; 3) the inferior pulmonary venous confluence; 4) the halfway between levels 3 and 5; 5) immediately above the right hemidiaphragm dome; 6) 2-4 cm above the posterior costophrenic angle (basal lungs). The halfway between levels 3 and 5 can be selected based on the image number or the coronal CT images. Due to anatomical variations (e.g., diaphragmatic eventration, pulmonary vein variations), ILD severity (with severe lower-lobe volume loss), and technical issues, the levels (especially levels 4-6) chosen can be slightly different from the predefined anatomical levels and should be appropriately adjusted.
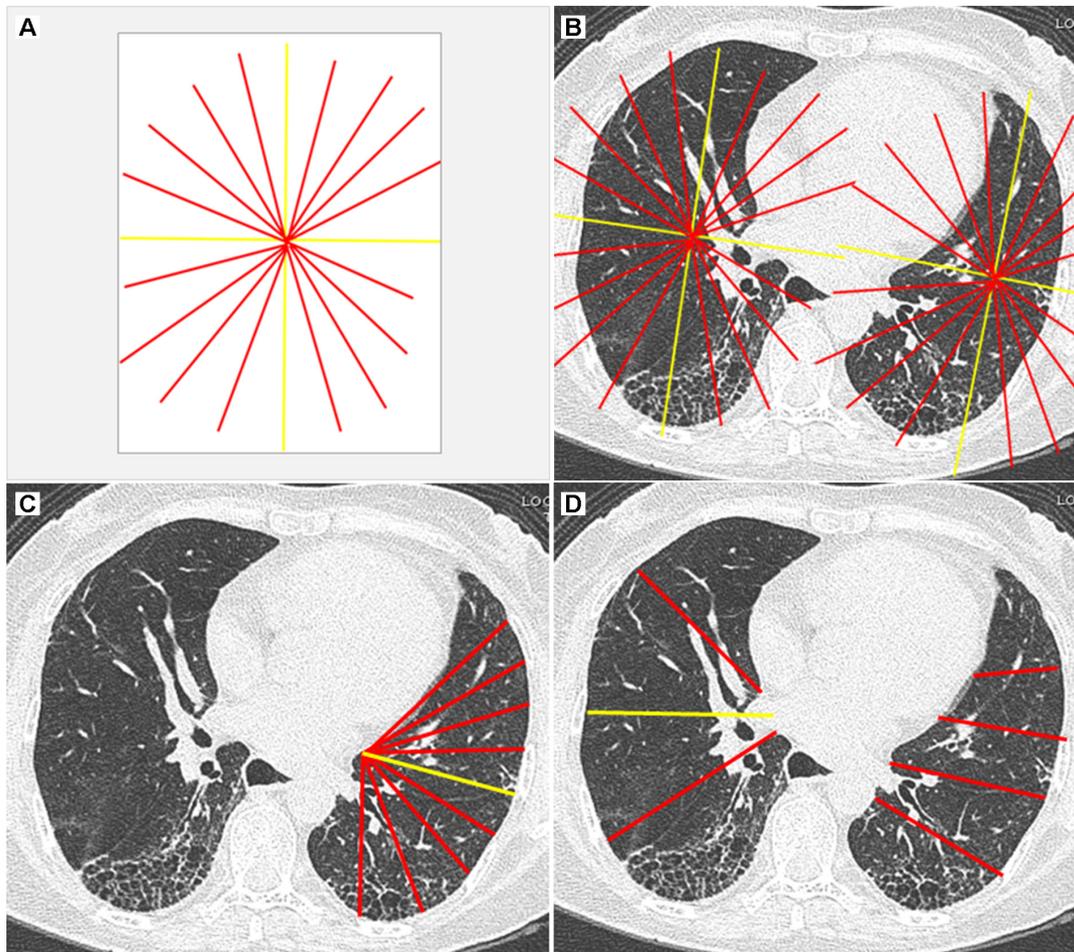
The extent of pulmonary involvement in the twelve zones (six on the right and the other six on the left) is then estimated in percentage to the nearest 5%, i.e., 0%, 5%, 10%, 15%, …,95%, 100%. A scoring tool (A and B) or arbitrary lines can be applied to aid a more precise estimation (Figure 2).

According to this method, the global disease extent and the fibrotic extent are derived from the summed percentages (extent) of these abnormalities in the six predefined levels or 12 lung zones divided by 12 (Tables 1 and 2). If level 5 is close to or similar to level 4 or 6, level 5 can be skipped. In this regard, the global disease extent and the fibrotic extent will be divided by 10 instead of 12. The global disease extent and the fibrotic extent can be reported in range, e.g., 20%-25% or 30%-35% of lung parenchyma (Tables 1 and 2).

Hence, the global disease and the fibrotic extent can be similar or different (Figure 1, Tables 1 and 2). In a patient with an inactive disease with irreversible fibrosis, the global disease and the fibrotic extent can be equal. However, the global disease extent is usually greater than the fibrotic extent in a patient having areas of pure ground-glass opacity or consolidation reflecting active interstitial inflammation or organizing pneumonia [10].

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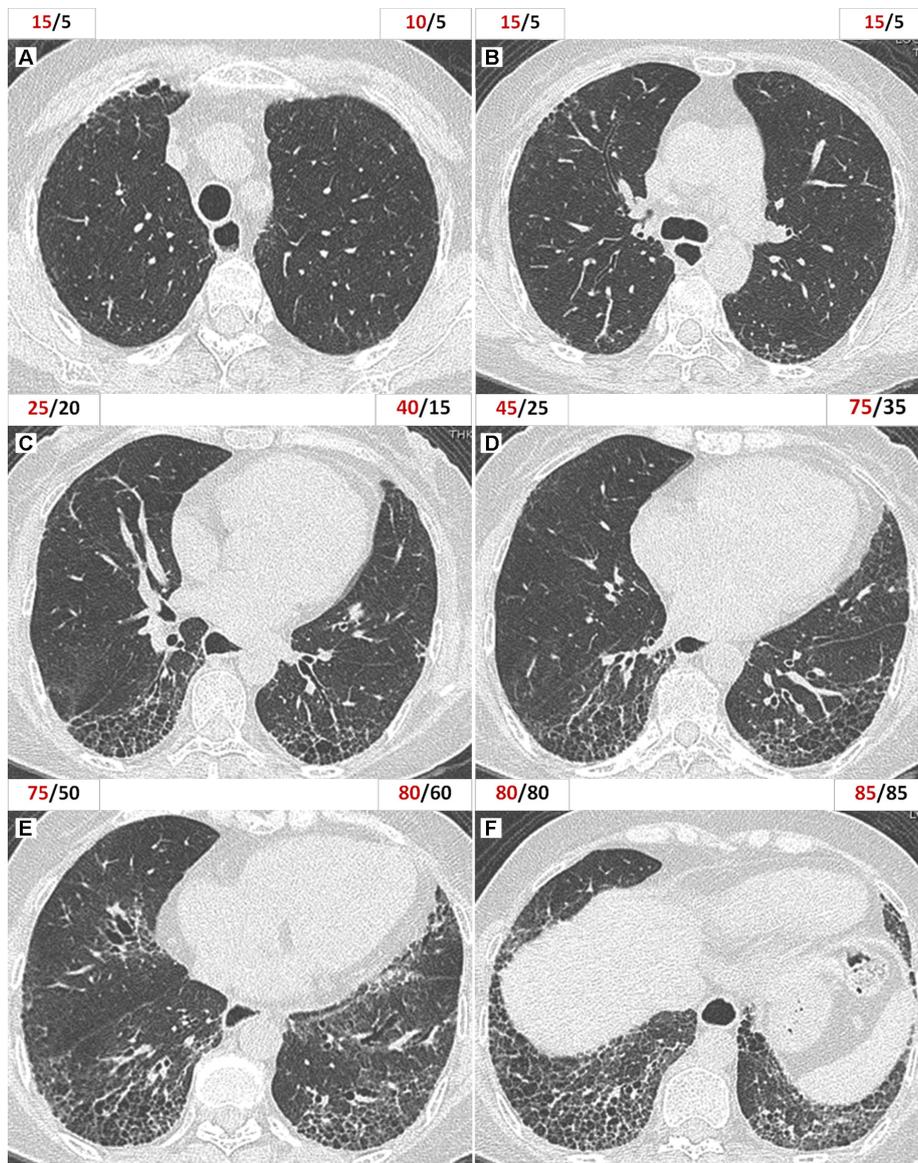
Figure 1. *The six predefined levels of HRCT images of a 58-year-old woman with systemic sclerosis -associated ILD, including A) level 1 (the origin of the great vessels, immediately above the top of the aortic arch), B) level 2 (the main carina), C) level 3 (the inferior pulmonary venous confluence), D) level 4 (the halfway between levels 3 and 5), E) level 5 (immediately above the right hemidiaphragm dome, and F) level 6 (2-4 cm above the posterior costophrenic angle). Each level consists of two lung zones. Pulmonary involvement in the twelve zones is estimated to be the nearest 5%. The red-colored number in each box denotes the estimated percentage of the global disease extent, whereas the black-colored number denotes the estimated percentage of the fibrotic extent.*

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Figure 2. *The illustrative images showing a scoring tool (A and B) and arbitrary lines (C and D) drawn on the axial HRCT images help quantify the percentages of the global disease extent and the fibrotic extent in the right and left lungs at level 3.*

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Table 1. *Example for calculating the global disease extent of the illustrative case in figure 1.*

The six predefined levels	Right lung (%)	Left lung (%)	The summed extent (%) of both lungs
1. Origin of the great vessels	15	10	25
2. Main carina	15	15	30
3. Inferior pulmonary venous confluence	25	40	65
4. Halfway between level 3 and level 5	45	75	120
5. Immediately above the right hemidiaphragm dome	75	80	155
6. 2-4 cm above the posterior costophrenic angles	80	85	165
Summed % of all 6 levels or 12 areas)			560
The global disease extent (summed % divided by 12)			46.7*

The global disease extent can be reported as 45%-50%.

Table 2. *Example for calculating the fibrotic extent of the illustrative case in figure 1.*

The six predefined levels	Right lung (%)	Left lung (%)	The summed extent (%) of both lungs
1. Origin of the great vessels	5	5	10
2. Main carina	5	5	10
3. Inferior pulmonary venous confluence	20	15	35
4. Halfway between level 3 and level 5	25	35	60
5. Immediately above the right hemidiaphragm dome	50	60	110
6. 2-4 cm above the posterior costophrenic angles	80	85	165
Summed % of all 6 levels or 12 areas)			390
The fibrotic extent (summed % divided by 12)			32.5*

The fibrotic extent can be reported as 30%-35%.

It is important to note that this proposed visual scoring system roughly estimates the extent of pulmonary involvement. It does not directly determine the severity based on morphological changes, for example, coarseness of reticular interstitial abnormalities, traction bronchiolectasis, traction bronchiectasis, and honeycombing. Hence, it cannot replace standard HRCT reporting and should be interpreted along with other HRCT features. Nevertheless, this proposed visual scoring method still serves as a complementary tool for initial evaluation, monitoring progression and treatment efficacy of ILD, and facilitating communication with clinicians.

Conclusion

The newly proposed visual scoring method for quantifying the global disease extent and the fibrotic extent on HRCT made by the consensus of the multidisciplinary working group for ILD in Thailand deems reproducible and straightforward. It is potentially helpful for managing ILD patients in monitoring disease progression, response to therapy, and eligibility for novel antifibrotic therapies.

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