

SUPPLEMENTARY FILES

Supplementary file 1: Details on CFCT score, as provided by the corelab LungAnalysis, Rotterdam, the Netherlands

Every lobe was scored for bronchiectasis, airway wall thickening, mucous plugging, parenchym abnormalities on inspiratory images and air trapping on expiratory images (if available). Each parameter was scored from 0 to 3, according to the table. The total CFCT score per lobe is the sum of all the subscores, the total CFCT score is the sum of the scores for all 6 lobes. If a lobe is resected or shows complete atelectasis, only maximum score (3) for consolidation can be counted for that lobe. Bronchiectasis, airway wall thickening and mucus plugging should be scored as 0 for that lobe. Only 2 patients with lobectomy were included in this study.

CT abnormality	Score			
	0	1	2	3
1. Bronchiectasis				
Central lung (extent of lung)	Absent	<33%	33%-67%	>67%
Peripheral lung (extent of lung)	Absent	<33%	33%-67%	>67%
Size of the largest	Absent	B<2V	B=2-3V	B>3V
Size of the average	Absent	B<2V	B=2-3V	B>3V
2. Airway wall thickening				
Severity	Absent	33%-50% V	50%-100% V	>100% V
Central lung (extent of lung)	Absent	<33%	33%-67%	>67%
Peripheral lung (extent of lung)	Absent	<33%	33%-67%	>67%
3. Mucous plugging				
Large airways (extent)	Absent	<33%	33%-67%	>67%
Small airways (extent)	Absent	<33%	33%-67%	>67%
4. Parenchyma				
Atelectasis and consolidation (extent)	Absent	<33%	33%-67%	>67%
Bulla and cysts (extent)	Absent	<33%	33%-67%	>67%
Ground glass opacification (extent)	Absent	<33%	33%-67%	>67%
5. Air trapping				
Extent	Absent	<33%	33%-67%	>67%
Pattern	Absent	Subsegmental	Segmental	Lobar

B: bronchial lumen diameter

V: vessel diameter

The CFCT subscores were then calculated per lobe as followed:

- Bronchiectasis score (range 0-12):

= (extent of bronchiectasis in central lung + extent of bronchiectasis in peripheral lung) x average bronchiectasis size multiplier

Where average bronchiectasis size = (size of the largest dilated bronchus + average size of dilated bronchi) / 2

And multiplier depends on average bronchiectasis size:

Average bronchiectasis size	Multiplier
0.5	0
1	1
1.5	1.25
2.0	1.5
2.5	1.75
3	2

- Bronchial wall thickening score (range 0-9)

= (extent of bronchial wall thickening in central lung + extent of bronchial wall thickening in peripheral lung) x severity of bronchial wall thickening multiplier

Bronchial wall thickening severity score	Multiplier
1	1
2	1.25
3	1.5

- Mucous plugging score (range 0-6)

= extent of mucous plugging in central lung + extent of mucous plugging in peripheral lung

- Parenchyma score (range 0-9)

= extent of dense parenchymal opacity + extent of ground glass opacity + extent of cysts or bullae

- Air trapping score (range 0-4.5)

= extent of air trapping x pattern of air trapping multiplier

Air trapping pattern score	Multiplier
1	1
2	1.5

Supplementary file 2: Intra rater variability and inter rater variability correlation coefficients, based on the reinterpretation of a subset of 5 chest CT's.

	Intrarater variability	Interrater variability
Bronchiectasis	0.822	0.728
Airway wall thickening	0.870	0.640
Mucus plugging	0.806	0.589
Parenchymal consolidation	0.710	0.847
Air Trapping (n=24)	0.369	0.251
Total CF-CT score	0.853	0.880

Supplementary file 3: Normal values for LCI, Scond*VT and Sacin*VT (c) as reported by Houltz[27] and Boon. The results are expressed as mean +/-SD.

	Houltz healthy controls 7-19 years	Houltz healthy controls 20-39 years	Boon Healthy controls 5-25 years
LCI	6.54 ± 0.28	6.70 ± 0.36	7.03 ± 0.68
Scond*VT	0.021 ± 0.004	0.021 ± 0.008	0.013 ± 0.009
Sacin*VT	0.051 ± 0.012	0.056 ± 0.021	0.053 ± 0.036

Supplementary file 4: Results for total HRCT score and subscores, absolute values and %. Results are expressed as median (IQR) and range. Abnormal scores are defined as a % >5% for that (sub)score.

	Absolute value	% of theoretical maximum score	Number of patients with abnormal score
Bronchiectasis	4.5 (3.0 – 7.4)	6.3 (4.2 – 10.3) Range 1.4 – 28.8	21 (70%)
Airway wall thickening	5.0 (3.0 – 7.8)	13.9 (8.3 – 21.9) Range 5.6 – 49.3	29 (97%)
Mucus plugging	2.0 (1.0 – 4.8)	3.7 (1.9 – 8.8) Range 0 – 16.7	14 (47%)
Parenchymal consolidation	3.0 (1.0 – 4.0)	5.6 (1.9 – 7.4) Range 0 – 13.0	15 (50%)
Air Trapping (n=24)	1.0 (0.0 – 3.0)	3.7 (0.0 – 11.1) Range 0 – 29.6	11 (45.8%)
Total CF-CT score	17.4 (10.6 – 25.2)	8.4 (3.1 – 12.2) Range 0.9 – 26.8	22 (73%)