

## **Online supplement**

### **Scoring methods for the area of fibrosis and low attenuation area on HRCT**

A thoracic radiologist with 28 years of experience re-read the chest HRCT, blinded to the clinical course and examination data for each patient. The lungs were divided into six zones (upper, middle, and lower zones in both lungs), and each zone was evaluated separately. The upper zone was defined as the part of the lung above the level of the tracheal carina; the lower zone as the part of the lung below the level of the inferior pulmonary vein; and the middle zone as the portion of the lung between the upper and lower zones. The extent of involvement of the findings was evaluated visually and independently for each lung zone. A score was assigned on the basis of the percentage of lung parenchyma that showed evidence of abnormality and was estimated to the nearest 10% of parenchymal involvement. The overall percentage of involvement was calculated by averaging the scores of the six lung zones [1].

Table S1. Prediction of elevated MPAP with continuous variables and binary variables.

Variable	OR (95% CI)	p value	Variable	OR (95% CI)	p value
SpO <sub>2</sub> , %	0.703 (0.612-0.807)	<0.001	SpO <sub>2</sub> ≥ 96%	0.235 (0.125-0.441)	<0.001
PaO <sub>2</sub> , mmHg	0.938 (0.914-0.962)	<0.001	PaO <sub>2</sub> ≥ 80 mmHg	0.255 (0.139-0.466)	<0.001
FVC, % pred	0.980 (0.966-0.995)	0.009	FVC ≥ 75%	0.380 (0.207-0.698)	0.002
DLco, % pred	0.952 (0.934-0.969)	<0.001	DLco ≥ 50%	0.174 (0.094-0.321)	<0.001
MMRC	1.986 (1.486-2.655)	<0.001	MMRC ≥ 2	3.313 (1.844-5.952)	<0.001
6MWD, m	0.996 (0.993-0.998)	<0.001	6MWD ≥ 540 m	0.405 (0.227-0.722)	0.002
Lowest SpO <sub>2</sub> , %	0.925 (0.897-0.955)	<0.001	Lowest SpO <sub>2</sub> ≥ 80%	0.259 (0.143-0.469)	<0.001
PA:Ao ratio	1.889 (1.466-2.434)	<0.001	PA:Ao ratio (/ 0.1) ≥ 9.0	3.745 (2.054-6.826)	<0.001
LAA, %	1.052 (1.018-1.086)	0.002	CPFE (LAA ≥ 10% in upper zone)	2.025 (1.088-3.767)	0.03
Estimated RVP, mmHg	1.019 (1.002-1.037)	0.03	Estimated RVP ≥ 30 mmHg	1.398 (0.785-2.492)	0.26

MPAP, mean pulmonary arterial pressure; PaO<sub>2</sub>, partial pressure for oxygen; FVC % pred, percent predicted forced vital capacity; DL<sub>CO</sub> % pred, percent predicted diffusion capacity for carbon monoxide; MMRC, modified Medical Research Council dyspnoea scale; 6MWD, 6-min walk distance; PA:Ao ratio, pulmonary artery diameter (PA) to ascending aorta diameter (Ao)

ratio on chest computed tomography; LAA, low attenuation area; CPFE, combined pulmonary fibrosis and emphysema; RVSP, right ventricular systolic pressure.

Table S2. Inter-rater reliability of CT findings.

Variable	ICC (95% CI)	p value
PA	0.852 (0.726-0.910)	<0.001
Ao	0.919 (0.888-0.940)	<0.001
PA:Ao	0.856 (0.803-0.893)	<0.001
PA:Ao $\geq$ 0.9	0.706*	<0.001

\*, Cohen kappa

ICC, intraclass correlation coefficient; CI, confidence interval; PA, pulmonary artery diameter; Ao, ascending aorta diameter.

## Reference

1. Sumikawa H, Johkoh T, Ichikado K, Taniguchi H, Kondoh Y, Fujimoto K, Tateishi U, Hiramatsu T, Inoue A, Natsag J, Ikemoto M, Mihara N, Honda O, Tomiyama N, Hamada S, Nakamura H, Muller NL. Usual interstitial pneumonia and chronic idiopathic interstitial pneumonia: analysis of CT appearance in 92 patients. *Radiology* 2006; 241: 258-266.