Development of prediction scoring system of acute exacerbation in idiopathic pulmonary fibrosis

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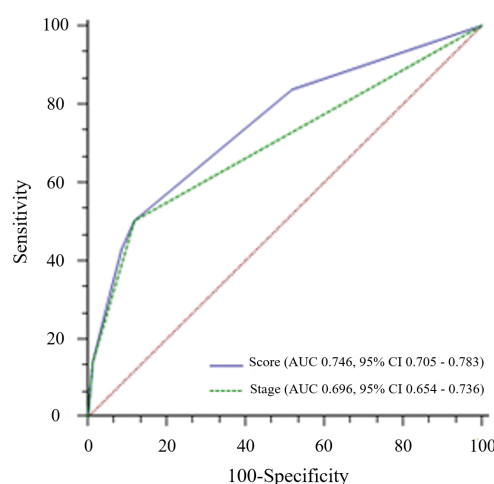
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Background/Aims: Idiopathic pulmonary fibrosis (IPF) is a chronic progressive fibrosing interstitial lung disease (ILD) with the worst prognosis. Acute exacerbation (AE) of IPF has the most disastrous impact on prognosis as a major cause of morbidity and mortality. However, the occurrence of AE is unpredictable and there is no proven treatment. Therefore, our aim in this study was to evaluate the risk factors for AE in patients with IPF in the Korea IPF Cohort (KICO) registry and to develop a prediction scoring system for AE.

Methods: This is a retrospective study of Korean patients with IPF in the KICO registry from June 2016 to February 2022. We developed a prediction scoring system for AE in the KICO registry.

Results: Among 707 patients in the KICO registry, 678 patients were enrolled. Male (82.0%) was predominant, and the mean age was 69.4 years. AE occurred in 165 patients (24.3%) during follow-up (median: 40.7 months). The mean time from IPF diagnosis to AE was 15.3 months. The lower FVC, shorter distance during 6MWT, and use of home oxygen were independently associated with AE in the multivariable logistic analysis. The mortality rate of AE-IPF was 36.4% (95% CI: 0.294-0.439). In the multivariable Cox analysis, older age was only an independent risk factor for mortality in IPF patients with AE. In risk predicting model using variables of FVC, distance during 6MWT, and use of home oxygen, there was a significant difference for predicting AE between the score (AUC − 0.746, 95% CI: 0.705-0.783, $p < 0.001$) and stage (AUC − 0.696, 95% CI: 0.654-0.736, $p < 0.001$), respectively.

Conclusions: The incidence and mortality rate of AE in patients with IPF in the KICO registry was 24.3% and 36.4%, respectively. We developed the prediction model for AE using variables including the use of home oxygen, distance during 6MWT, and FVC.

Variable	Score	No. of patients (%)
Home oxygen		
Yes	3	423(83.4)
No	0	84(16.6)
FVC, % predicted		
>75	0	233(46.0)
50-75	1	247(48.7)
<50	2	27(5.3)
Distance, m		
≥250	0	457(90.1)
<250	1	50(9.9)
Score		
0	0	200(39.4)
1	1	194(38.3)
2	2	23(4.5)
3	3	29(5.7)
4	4	36(7.1)
5	5	18(3.6)
6	6	7(1.4)
Stage		
I	0-1	394(77.7)
II	2-4	88(17.4)
III	5-6	25(4.9)

Laboratory Evaluation	Cut-Point Value	Group AE (+)	Group AE (-)	Cut-Point Value	AUC (p)	Sensitivity , %	Specificity , %	PPV , %	NPV , %
Score	>1	68	45	>1	.746* (<.001)	50.4	87.9	60.2	83.0
	≤1	67	327						
Stage	>1	68	45	>1	.696* (<.001)	50.4	87.9	60.2	83.0
	≤1	67	327						