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POS1289 MACHINE LEARNING ALGORITHM AS A USEFUL TOOL IN THE GREY AREA OF CARDIOPULMONARY MORTALITY IN SYSTEMIC SCLEROSIS

Keywords: Artificial intelligence, Cardiovascular disease, Systemic sclerosis

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Background: Prognosis of systemic sclerosis (SSc) patients is related to the presence of major internal organ involvement and specific autoantibody positivity. Cardiopulmonary complications are the leading cause of death in patients with SSc.
Objectives: The objective of the study is to evaluate the ability of an artificial intelligence algorithm based on laboratory and functional variables associated to SSc cardio-pulmonary involvement to predict the 5-year related mortality.
Methods: Laboratory and functional parameters of cardiopulmonary involvement of SSc patients symptomatic for dyspnoea, palpitations, or chest pain were recorded. Forced vital capacity (FVC), alveolar diffusion of CO (DLco), serum troponin, NT-pro-BNP, cardiac ejection fraction (EF) and pulmonary arterial systolic pressure (PAPs) were used for clustering by a partition around medoids (PAM) algorithm. The resulting clusters were compared for 5-year mortality due to SSc cardio-pulmonary causes.
Results: 216 patients were enrolled, aged 54.5±14.4 years, and with a disease duration of 7.4±7.0 years. The 12% of patients were male, 41% had a diffuse cutaneous disease, 45% were anti-Scl70 positive. With specific reference to cardiopulmonary involvement, 50% of patients had pulmonary fibrosis at HRCT, 15% pulmonary arterial hypertension, 16% a EF <55%, 11% increased levels of troponin, and 42% elevated NT-proBNP levels. During follow-up, 26 patients (12%) died because of causes related to cardiac-pulmonary involvement. The machine learning algorithm identified two different groups according to prognosis ($\chi 2(2) = 29.76$, $p<0.001$) with a 5-year mortality of 15% and 0.8%. Patients with higher mortality were characterized by lower FVC values and higher NT-proBNP and troponins compared to rest of the cohort ($p<0.001$). Furthermore, the prevalence of male gender, diffuse cutaneous disease, anti-Scl70 positivity and anti-centromere negativity were higher in the high-risk group compared with the alternative cluster ($p<0.05$ for all characteristics). Interstingly, in high mortality cluster a relevant percentage of patients apparently do not present classical risk factors for progressive SSc, in fact 40% were females, 30% of patients presented a limited cutaneous involvement, 36% were anti-Scl70 negative and 30% were anti-centromere positive, 26.4% did not presented pulmonary fibrosis, and 35% of patients did not have pulmonary hypertension.
Conclusion: An artificial intelligence algorithm based on laboratory-functional parameters of SSc cardio-pulmonary involvement was shown to be able to predict the prognosis of SSc patient more accurately than traditional predictors. Given the high mortality risk of patients with SSc, the application of dedicated prognostic algorithms could better support the clinician in the management of patients.
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POS1290 INCREASED RISK OF MYOCARDIAL INFARCTION AND STROKE IN PATIENTS WITH SYSTEMIC SCLEROSIS: A NATIONWIDE COHORT STUDY

Keywords: Epidemiology, Systemic sclerosis, Cardiovascular disease

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Background: Previous studies have suggested a link between systemic sclerosis (SSc) and cardiovascular disease, but large-scale data are still lacking due to the nature of rare autoimmune diseases.
Objectives: We aimed to compare the incidence of myocardial infarction (MI) and stroke in patients with SSc and age- and sex-matched controls in a nationwide population-based cohort in Korea.
Methods: We included patients with SSc defined by the ICD-10 code (M34) and rare and intractable disease code (V138) and 1:5 age- and sex-matched controls using the Korean National Health Insurance Database. The outcomes of the study were MI and stroke. Cox proportional hazard analysis and Kaplan-Meier curve were used to compare the incidence of outcomes between patients with SSc and controls.
Results: A total of 4700 patients with SSc and 23500 controls were included in the study. The mean follow-up period was 5.6 ± 2.8 years. At baseline, patients with SSc had higher prevalent rates of comorbidities such as hypertension, hyperlipidemia, and congestive heart failure than controls. Patients with SSc had a 3-fold higher risk of MI (adjusted hazard ratio [aHR] 3.01, 95% confidence interval [CI] 2.41–3.76) and a 1.7-fold higher risk of stroke (aHR 1.65, 95% CI 1.28–2.14) compared to controls. There were no differences in the association between SSc and MI or stroke by age, sex, or comorbidities.
Conclusion: This nationwide population-based cohort study revealed an association between SSc and increased risk of MI and stroke. Therefore, careful monitoring and preventive measure for the cardiovascular diseases in patients with SSc are required.
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POS1291 PREDICTION OF MORTALITY IN SSC-ILD DEPENDS ON DEFINITION OF ILD PROGRESSION

Keywords: Lungs, Systemic sclerosis, Prognostic factors

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Background: Progression of interstitial lung disease (ILD) is a candidate for long-term mortality in patients with systemic sclerosis (SSc). Different definitions of progression have been proposed. Declining lung function is often used, whereas others include composite definitions such as the 2022 ATS/ERS/JRS/ALAT guideline criteria for progressive pulmonary fibrosis (PPF) and the INBUILD criteria for progressive fibrosing ILD (PF-ILD). These different definitions have not been compared in SSc-ILD.
Objectives: To estimate the prevalence of ILD progression applying different definitions and test their performance of predicting mortality.
Methods: We included all SSc patients from the Oslo and Zurich cohorts who had ILD on HRCT and serial assessments of disease progression defined as:
(A) Absolute FVC decline >5% over 12 months
(B) PPF guideline criteria with 2/3 criteria present over 12 months of (1) worsening of respiratory symptoms; (2) absolute decline in FVC >5% or in DLCO >10% and (3) disease progression on HRCT
(C) INBUILD PF-ILD criteria within 24 months with (1) FVC decline ≥10%, (2) FVC decline >5-<10% and worsening of respiratory symptoms or increased lung fibrosis on HRCT, or (3) worsening of respiratory symptoms and increased lung fibrosis.
We assessed the prevalence of ILD progression using these competing definitions and applied multivariable cox regression models (with hazards ratios (HR) and 95%CI) adjusted for known risk factors for mortality and compared the performance using Harrels c-index.
Results: In total, 231 SSc-ILD patients from Oslo and Zurich were included, with 71 (31%) showing FVC decline >5%, 43 (19%) fulfilling the PPF guideline and 89 (39%) the INBUILD PF-ILD criteria. Most progressive patients fulfilled ≥1 definition of progression [60/107 (56%)] while 124 (54%) did not progress (Figure 1). Patient characteristics did not differ between the definitions, except for more extensive ILD and ground glass on HRCT and more frequent oxygen desaturation among those fulfilling the PPF criteria (Table 1). The number of deaths [44 (47%)] over mean 7.7 years (SD 3.9) follow up were comparable in the different groups. The progression definitions performed differently in multivariable cox models adjusted for age, sex, disease duration, SSc subtype, extent of lung fibrosis, baseline FVC and treatment using FVC decline>5% (HR1.87, 1.10-3.17

95%CI; p=0.020; c-index 0.7331), PPF guideline (HR1.42, 0.79-1.84 95%CI; p=0.231; c-index=0.7156) and INBUILD PF-ILD (HR 2.38, 1.40-4.04 95%CI: p<0.001; c-index=0.7338). The models discriminating ability was not significantly different (p=0.138).

Conclusion: The prevalence of ILD progression varies depending on which definition was applied. FVC decline alone and PF-ILD criteria predicted mortality significantly but not the 2022 PPF guideline criteria.

	FVC decline>5%	PPF	PF-ILD
Male sex, n (%)	20 (28)	12 (28)	25 (28)
Age at onset, y (SD)	49 (14.8)	50 (13.3)	49 (14.8)
Disease duration<3y, n (%)	43 (61)	24 (56)	33 (38)
dcSSc, n (%)	38 (54)	24 (56)	47 (53)
ATA, n (%)	33 (47)	20 (47)	35 (39)
CRP↑, n (%)	7 (14)	4 (15)	7 (14)
FVC, % (SD)	93 (19.6)	89 (19.5)	92 (20.7)
DLCO, % (SD)	66 (16.3)	61 (16.2)	64 (17.8)
Functional class 3&4, n (%)	9 (19)	6 (19)	12 (19)
ILD>10%, n (%)	20 (43)	18 (56)	26 (47)
Ground glass, n (%)	28 (42)	23 (56)	42 (48)
O_2 desaturation, n (%)	6 (14)	7 (30)	10 (18)
Immunosuppressives, n (%)	23 (32)	21 (49)	34 (38)
Death, n (%)	32 (45)	20 (47)	39 (44)

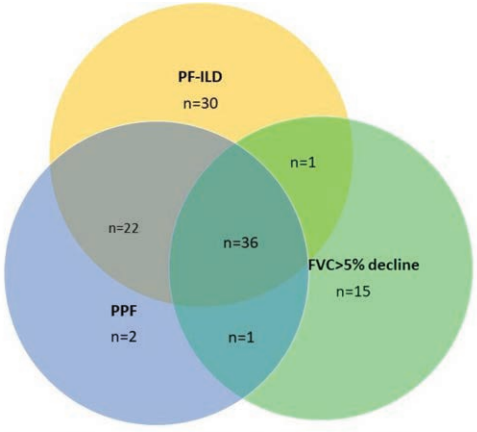

Figure 1: Venn diagram of patients fulfilling the different definitions of ILD progression

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POS1292 **WITHDRAWN**