established protocols. Clinical charts were used to analyze clinical and laboratory data for possible statistical correlations, performed by Prism (GraphPad Software 4.0, Fisher exact test) with statistical significance set at p values <0.05 .

Results: We identified a new protein-IP pattern characterized by a set of several proteins of 140/40-25kD in 8/63 (13%) SSc sera (Figure panel A). This IP pattern corresponds to the hnRNP complex which is composed of several proteins and RNAs involved in RNA processing and splicing. These proteins have molecular weights ranging between 32 and 45kD as the set of proteins we identified in our experiments. To define the 140/40-25kD pattern identified in 8 SSc cases, we performed IP-WB using two anti-hnRNP antibodies selected by the molecular weight of the target antigen. We tested anti-hnRNP C1+C2 and anti-hnRNP L monoclonal antibodies in these 8 samples after preparation by crosslinking of IgG to PAS beads and protein-IP protocol, followed by WB (Figure panel B). Results show that 5/8 SSc anti-140/40-25kD samples are positive for hnRNP L, while no reactivity was observed with hnRNP C1+C2 monoclonal antibody. We further observed that anti-hnRNP antibodies are significantly associated with serum ACA positivity ($p=0.008$), severe Raynaud's phenomenon with digital ulcers requiring IV prostacyclin (75%, 6/8 vs 7/54 ACA+, 13%; $p=0.0006$), and esophageal involvement in 4 cases.

Conclusion: We identified anti-hnRNP antibodies as new target antigen of a subset of SSc patients with ACA positivity and severe peripheral vascular involvement. Additional analysis is necessary to study the several components of anti-hnRNP antibodies, and longitudinal data will provide evidence on the predictive value of this biomarker.

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SAT0268

CLINICAL FEATURES OF INTERSTITIAL LUNG DISEASE ASSOCIATED WITH KOREAN IDIOPATHIC INFLAMMATORY MYOPATHIES ACCORDING TO THE AUTOANTIBODY PROFILE

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Background: Myositis-specific and myositis-associated autoantibodies (MSA and MAA) have been used for more detailed classification and prediction

Figure 1. Prevalence of MSA/MAA in IIM patinetns with ILD (N=43)

Table 1. Clinical features and autoantibody profile of IIM-ILD patients

	IIM-ILD (n=43)	IIM-ILD (n=43)
Age (years)*	55.6±12.0	Myositis-Specific Autoantibodies (MSA)
Gender (F:M)	33 : 10	Anti-ARS 20 (46.5)
Smoking Hx	5 (11.6)	Anti-Jo-1 10 (23.3)
Raynaud phenomenon	9 (20.9)	OJ,EJ,PL7,PL12 11 (25.6)
Time of ILD onset		Anti-SRP 8 (18.6)
ILD preceding	7 (16.3)	Anti-MDA5 12 (27.9)
Concomitant with myositis	31 (72.1)	Anti-Mi2 4 (9.3)
myositis preceding	6 (14.0)	Anti-TIF1-r 1 (2.3)
HRCT Findings		Anti-SAE 1 (2.3)
NSIP	19 (44.2)	Myositis-Associated Autoantibodies (MAAs)
UIP	6 (14.0)	Anti-Ro52 26 (60.5)
COP	18 (41.9)	Anti-Ku 1 (2.3)
Pulmonary function test		
FEV1/FVC, %	85.0±13.7	
DLCO, %	64.8±23.2	
DLCO <65%	16 (37.2)	

of clinical course of idiopathic inflammatory myopathies (IIM). Interstitial lung disease (ILD) is known to be complicated with approximately 50% of IIM patients, having poor prognosis.

Objectives: The aim of this study is to investigate clinical features of ILD in Korean adult patients with IIM according to the autoantibody profile including MSA and MAA.

Methods: We conducted a multicenter cohort study including 108 adult patients (age ≥ 18 years) who have been diagnosed as IIM by ENMC criteria. MSA and MAA were screened with Immunoblot assay using Euro-line strip (EUROIMMUN, Germany). Clinical information including the time of developing ILD and imaging findings of chest CT was reviewed using by medical record.

Results: ILD was found in about half (52.8%, 57 of 108) of enrolled patients and closely associated with the presence of anti-ARS Abs, anti-MDA5 and anti-Ro52. Among 43 patients, Anti-ARS Abs were most common (46.5%) and included anti-Jo-1 (23.3%), -OJ (2.3%), -EJ (9.3%), -PL7 (14%), and -PL12 (7%) (Figure 1). To analyze the correlation of autoantibodies with pattern of ILD onset, we stratified the 43 patients into three groups: into ILD-preceding (7/43, 16.3%), simultaneous (31/43, 72.1%) and myopathy-preceding (6/43, 14%). In the ILD-preceding group, anti-ARS antibodies were frequent. ILD preceded myopathy by 19.5 months on average. The imaging pattern of ILD in the ILD-preceding group were Cryptogenic Organizing Pneumonia (COP) 6, Nonspecific Interstitial Pneumonia (NSIP) 1 and no Usual interstitial pneumonia (UIP). A half of patient in ILD-preceding group had Raynaud's phenomenon, and one patient had mechanic hand. On the other hand, in myopathy-preceding group, 30% of patients (2/6) were UIP and a half of patients had pulmonary function which required treatment (DLCO <65%).

Conclusion: In our study, ILDs were accompanied by over half of the cases of IIM and most of them had autoantibodies. There are characteristic clinical features by onset time of ILD, so paying attention to them will help early diagnosis and prognosis prediction of ILDs.

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SAT0269

THE IMPACT OF SMOKING ON NAILFOLD MICROANGIOPATHY AND AUTOANTIBODY STATUS IN MALE AND FEMALE PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Smoking is a risk factor for primary Raynaud's phenomenon and, in patients with systemic sclerosis (SSc), has detrimental effects on the intensity of peripheral microangiopathy⁽¹⁾. Two studies on SSc observed a lower prevalence of anti-topoisomerase I antibodies (ATA) in ever-smokers compared to never smokers^(2,3), but little is known about how the effect of smoking on autoantibody status differs in male and female patients.

Objectives: The aim of our study was to evaluate, in a cohort of SSc patients, the influence of smoking on nailfold videocapillaroscopy (NVC) patterns and autoantibody positivity, comparing findings between men and women.

Methods: We included 361 adult patients (279 women and 82 men) with SSc. At baseline NVC was performed and microvascular damage was qualitatively assessed based on normal findings, aspecific/secondary pattern, or distinct "early", "active", or "late" scleroderma patterns. Information about smoking status was collected, but data regarding duration and intensity were not available for all individuals, so patients were categorized as never smokers or ever smokers. The presence of SSc-specific ATA, anti-centromere (ACA), and anti-RNA polymerase III (ARA) antibodies was investigated at baseline. Chi-square was used to compare proportions and significance level was set at 0.05.

Results: No significant difference was observed in the qualitative assessment of microangiopathy between ever smokers and never smokers, nor in autoantibody positivity. Scleroderma patterns early, active, and late, were respectively present in 12%, 44% and 21% of ever-smokers, and 9%, 48%, and 29% of never-smokers. Equal proportions of NVC patterns were found also in the gender-based sub-cohorts. Regarding autoantibodies, 27% of women who never smoked were ATA positive compared to 11% of ever smokers ($p < 0.001$), while the opposite trend was observed in men, although not reaching statistical significance possibly due to the relatively small size of the male cohort.

Conclusion: In our large cohort of SSc patients, microangiopathy was not influenced by smoking status in male or female patients. The prevalence of ATA was, albeit not significantly, lower in ever smokers. Our finding is in accordance with previous studies^(2,3), but it is worth noting that this disparity is largely contributed by the statistically significant higher frequency of ATA positivity in women who never smoked, compared to ever smokers.

Table 1. microangiopathy pattern and autoantibody positivity in ever smokers and never smokers in all SSc patients and in gender-based sub-cohorts. ATA: anti-topoisomerase I, ACA: anti-centromere, ARA: anti-RNA polymerase III, NVC: nailfold videocapillaroscopy.

	All SSc patients			Men			Women		
	Ever smokers	Never smokers	p-value	Ever smokers	Never smokers	p-value	Ever smokers	Never smokers	p-value
n (%)	208 (58)	153 (42)		62 (76)	20 (24)		146 (52)	133 (48)	
NVC Pattern									
scleroderma early, n (%)	26 (12)	14 (9)	ns	5 (8)	0	ns	21 (14)	14 (10)	ns
scleroderma active, n (%)	91 (44)	73 (48)	ns	24 (39)	11 (55)	ns	67 (46)	62 (47)	ns
scleroderma late, n (%)	44 (21)	45 (29)	ns	13 (21)	5 (25)	ns	31 (21)	40 (30)	ns
normal, n (%)	4 (2)	0	ns	2 (3)	0	ns	2 (1)	0	ns
aspecific/secondary, n (%)	43 (21)	21 (14)	ns	15 (24)	3 (15)	ns	20 (14)	14 (10)	ns
Autoantibodies									
ATA, n (%)	41 (20)	40 (26)	ns	25 (40)	4 (20)	ns	16 (11)	36 (27)	<0.001
ACA, n (%)	90 (43)	66 (43)	ns	10 (16)	6 (30)	ns	80 (55)	60 (45)	ns
ARA, n (%)	11 (5)	5 (3)	ns	2 (3)	0	ns	9 (6)	5 (4)	ns

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SAT0270

COMPREHENSIVE ICF CORE SET AND ICF-BASED QUESTIONNAIRE ASSESSING ACTIVITIES AND PARTICIPATION IN PATIENTS WITH SYSTEMIC SCLEROSIS: THE SCISCF STUDY

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Background: Recent guidelines regarding systemic sclerosis (SSc) emphasized the need of measuring functioning along with organ-specific measures, pointing out the importance of patient-reported outcome measures (1). The International Classification of Functioning, Disability and Health (ICF) endorsed by the WHO in 2001 set a unified language covering all dimensions of human functioning, including body structures, body functions, activities and participation and environmental factors.

Objectives: To develop a comprehensive ICF core set for SSc and to conceive a patient-centered ICF-based questionnaire assessing activities and participation in patients with SSc.

Methods: The development of the ICF comprehensive core set followed 2 steps (2). In the first step, meaningful concepts related to SSc were collected using data source triangulation from patients (N=18), experts (N=10) and literature (N=174 articles). In the second step, concepts were linked to the best-matching ICF categories by 2 independent reviewers according to prespecified linking rules (3). Finally, patient-reported activities and participation categories of the comprehensive ICF core set were translated into understandable questions

Results: After linking concepts to ICF codes, 151 ICF categories were collected from focus groups, 22 from experts and 82 from literature. After fusion of the sources and removal of duplicates, the comprehensive ICF core set included 165 categories: 1 at the first level, 158 at the second level, 6 at the third level, with 50 categories on body functions, 15 on body structures, 53 on activities and participation and 47 on environmental factors. Patient-reported ICF categories on activities

Conclusion: We developed a comprehensive ICF core set that offers a conceptual framework for SSc patients' care and health policy. Using a patient-centered approach, we conceived a patient-centered ICF-based questionnaire, the Cochin Scleroderma ICF-65 questionnaire, assessing activities and participation in patients with SSc.

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