Table 2. Numbers and combinations of autoantibodies identified in the 96 SSc patients.

	Topo-I	CENP	RNAP III	Fibrillarin	NOR90	Th/To	Pm/Scl	Ku	PDGFR	Ro52
Topo-I	22	1	7	0	1	5	9	5	0	7
CENP		12	2	0	2	2	2	2	0	11
RNAPIII			4	0	1	2	1	3	0	7
Fibrillarin				0	0	0	0	0	0	0
NOR90					0	1	1	0	0	2
Th/To						0	4	2	0	2
Pm/Scl							4	3	0	6
Ku								1	0	2
PDGFR									0	0
Ro52										2
Single positive	22	12	4	0	0	0	4	1	0	2
Total	45	27	18	0	5	8	17	9	0	24

Figure 1. Diagram of disease-related antibodies against the four main autoantibodies [anti-centromere (antiCENP), anti-Topoisomerase I (antiTopo I), anti-RNA polymerase III (antiRNAP III) and anti-Ro52].

Conclusion: We presented the clinical and serologic features of the Turkish SSc patients from a new inception cohort. Clinical features of the SSc patients with single or multiple antibody positivity were not different.

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AB0619 PROGNOSTIC FACTORS OF PATIENTS WITH ANTI-MDA5 ANTIBODY-POSITIVE DERMATOMYOSITIS COMPLICATED WITH INTERSTITIAL PNEUMONIA -A JAPANESE SINGLE CENTER STUDY-

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Background: Anti-melanoma differentiation-associated gene 5 antibody (anti-MDA5 Ab)-positive dermatomyositis (DM) is frequently associated with rapidly progressive interstitial pneumonia (RPIP), whose prognosis is assumed to be poor[1]. Although outcome of DM-RPIP has been reported to be improved by early immunosuppressive therapy, we still experience the cases with severe outcome. Only several reports mentioned the prognostic factors and they have not been fully elucidated.

Objectives: To identify the predictors of prognosis in patients with anti-MDA5 Ab-positive DM associated with interstitial pneumonia (DM-IP).

Methods: Anti-MDA5 Ab-positive DM-IP patients admitted to Fujita Health University Hospital between January 2010 and October 2019 were consecutively included and stratified into 2 groups, the survived and the deceased groups. DM was diagnosed according to the criteria proposed by Bohan and Peter[2]. Clinically amyopathic DM was diagnosed according to the criteria proposed by Sontheimer [3]. Diagnosis of IP was based on findings of high resolution CT scan (HRCT). The definition of RPIP was rapid exacerbation of hypoxemia or HRCT findings in a period of days to one month after the onset. Clinical features and prognosis of the patients were collected retrospectively and compared between groups. Candidates of predictors are extracted by the univariable analysis using Fisher's exact test for dichotic parameters and Wilcoxon signed-rank test for

continuous parameters and multivariable analysis using logistic regression analysis. Receiver operating characteristic (ROC) curve analysis was examined to obtain the cut-off level. Survival was examined using Kaplan-Meier method and Log-rank test.

Results: Twenty-one patients were involved. Eight were deceased and 13 were survived. The deceased group had a higher ratio of male (75% versus 25%, $p=0.018$). All deceased cases were with RPIP and 67% in the survived cases. Levels of serum ferritin (4490 versus 646 ng/mL, $p=0.0026$), CRP (2.1 versus 0.9 mg/dL, $p=0.0490$), CK (1150 versus 290 U/L, $p=0.017$), AST (194 versus 108 U/L, $p=0.025$) and LDH (674 versus 368 U/L, $p=0.011$) were higher in the deceased group. Interestingly, skin ulcers were tended to be more frequent (12.5% versus 87.5%, $p=0.0587$), and anti-SS-A antibody was also more frequently detected (14.3% versus 85.7%, $p=0.0072$) in the survived group. Using ROC analysis cut-off values were 963 ng/mL for serum ferritin level (sensitivity 100%, specificity 83%), 0.7 mg/dL for CRP (sensitivity 75%, specificity 69%), 308 U/L for CK (sensitivity 88%, specificity 77%), 62 U/L for ALT (sensitivity 100%, specificity 62%), and 454 U/L for LDH (sensitivity 88%, specificity 77%). Patients were divided into two groups based on these cut-offs or based on dichotic parameters and survival was examined between 2 groups. Except CRP and anti-SS-A antibody, survival was significantly worse in parameter-positive or higher groups. Interestingly, anti-SS-A antibody-positive group had better outcome compared with those without.

Conclusion: In our analysis, novel candidates such as serum CK, AST, and LDH levels were newly extracted and parameters previously reported was also included and those were also associated with the clinical outcome. In addition, anti-SS-A antibody was identified as a novel protective factor associated with a good outcome.

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AB0620 EFFECTIVENESS OF RITUXIMAB IN PATIENTS WITH EARLY DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS. A MULTICENTER ANALYSIS.

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Background: Rituximab (RTX) is effective in improving skin affection in patients with diffuse cutaneous systemic sclerosis (DcSSc). However, there are few data on early use of this drug.

Objectives: To evaluate RTX effectiveness for skin disease in patients with DcSSc of less than 3 years of evolution.

Methods: Multicenter, observational and retrospective study. Patients with DcSSc starting RTX within 3 years since first non-Raynaud symptom were recruited. Demographic variables, time of disease duration at the beginning of RTX, immune pattern and time on RTX treatment were collected. Effectiveness was defined as modified Rodnan skin score (mRSS) improvement. Evaluations were done by the same experienced rheumatologist. Patients subjective perception of skin hardening and/or tightness was evaluated. mRSS changes from baseline to 6 and 12 months after RTX beginning and, later on, to the last available observation were analysed using Wilcoxon test. Statistical analysis was performed with SPSS 20.0.

Results: 11 patients (8 women) were recruited from 2 university hospitals. Median age was 48 years (IQR 22). Median time since diagnosis to RTX beginning was 12 months (IQR 8). 5, 3 and 2 patients presented ATA+, RNPIII+ and Ro-52+, respectively. Median duration of RTX treatment was 12 months (IQR 68). Median baseline mRSS was 15.5 (IQR 18). Median mRSS after 6 and 12 months of RTX treatment and at last available mRSS evaluation was 15 (IQR 13), 14.5 (IQR 13) and 11 (IQR 16), respectively. mRSS showed statistically significant improvement at 6 (29%, IQR 37) and 12 months of RTX treatment (35%, IQR 34) and, thereafter, at last available observation (39%, IQR 51), compared to basal mRSS. Most patients reported subjective improvement at 6 (9 of 10

patients) and 12 months (6 of 7), and at last available evaluation (6 of 8); all other patients reported stability.

Conclusion: In our experience, patients with DcSSc seem to benefit of early RTX treatment. Improvement may be seen as early as 6 months and seems to reach a plateau at 12 months.

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AB0621 TOLERABILITY AND SAFETY OF ACETYSALICYLIC ACID IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic Sclerosis (SSc) is characterized by an increased incidence of macro- and microvascular complications. Current evidences on efficacy, safety and tolerability of acetylsalicylic acid (ASA) in SSc patients are limited, and the indication to this treatment is based on the experience of each single centre or physician. Esophagus and stomach are the portions of the digestive tract that are more frequently affected by adverse events due to ASA exposure.

Objectives: We evaluated the incidence of adverse events associated with low-dose ASA treatment in a cohort of patients affected by SSc.

Methods: Demographic data and disease features of 302 patients affected by SSc treated with low-dose ASA were collected and patients were followed-up for a median period of 6.9 years (range: 0-20 years). The proportion of patients taking ASA for secondary prevention for cardiovascular disease was also noted. The incidence of discontinuation of the drug, gastrointestinal intolerance, bleeding and death in the observation period was recorded.

Results: Patients had a median age of 54.0 years (19.6-89.4); 91.9% were female, 13.2% were smokers and 44.0% had a BMI ≥ 30 Kg/m². The prevalence of ischemic heart disease, peripheral vascular disease and stroke was of 8.6%, 5.3% and 3.3%, respectively; 48.7% of the patient took ASA in primary cardiovascular prevention. Therapy started after a median disease duration of 4.8 years (range: 0.0- 30.1 years) since the first non-Raynaud symptom and 56.6% of patients had an early disease (less than three years of disease duration). During the observation period, 30 patients (14.3 per 1000 person-years) discontinued ASA after an average period of assumption of 4.6 years (range: 0.3-18.0 years). The main adverse events were heartburn, dyspepsia and hematochezia, recorded in 18 patients (8.6 per 1000 person-years). Eight of them (3.8 per 1000 person-years) had evidence of digestive tract bleeding. Five patients (2.4 per 1000 person-years) discontinued ASA due to recurrent epistaxis. Twenty-eight patients (13.4 per 1000 person-years) died in the follow-up period, 16 of these (7.6 per 1000 person-years) because of SSc-related causes. None of them had evidence of major bleeding. We used Kaplan-Meier analysis to evaluate the incidence of ASA discontinuation. The history of digital ulcers (Log rank test X^2 4.7, $p=0.037$) and male sex (Log rank test X^2 4.3, $p=0.03$) were associated with a higher cumulative ASA discontinuation rate due to gastrointestinal intolerance.

Conclusion: In our cohort of SSc patients, ASA resulted safe and well tolerated in most cases, despite the risk of gastroesophageal abnormalities due to disease. Although this comforting results, taking in account the lack of controlled-randomized trials about efficacy and safety, the choice to start antiplatelet therapy with ASA should be mandatorily preceded by a careful evaluation of risks and benefits. Furthermore, an attentive monitoring for possible adverse effects is needed during ASA treatment. Patients with digital ulcers and male sex could present less drug tolerability.

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AbbVie, Bristol-Myers Squibb, Celgene, Eli Lilly, Janssen, Merck Sharp & Dohme, Novartis, Sanofi, UCB, Roche, Pfizer, Speakers bureau: AbbVie, Bristol-Myers Squibb, Celgene, Eli Lilly, Janssen, Merck Sharp & Dohme, Novartis, Sanofi, UCB, Roche, Pfizer, Silvia Laura Bosello Speakers bureau: Abbvie, Pfizer, Boehringer

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AB0622 ASSOCIATIONS WITH DIGITAL PITTING IN SYSTEMIC SCLEROSIS: A RETROSPECTIVE ANALYSIS.

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Background: Digital pitting, the loss of tissue at the fingertip, is a cardinal feature of systemic sclerosis (SSc), contributing 3 of the 9 required points to fulfil the 2013 ACR/EULAR classification criteria. However, research into digital pitting has been scarce, despite it being painful and impacting on hand function.

Objectives: To identify factors associated with digital pitting in patients with SSc. **Methods:** This was a retrospective analysis of data from patients with SSc attending a tertiary referral centre. Patients were subdivided into those with and without digital pitting, as recorded at their last documented attendance. The following variables were analysed: age, gender, age at Raynaud's onset, age at SSc onset, limited/ diffuse cutaneous subtype, history of intravenous (IV) vasodilators, amputations, debridements and autoantibody status (anti-RNA polymerase, anti-Scl70, anti-centromere and anti-RNP).

Results: Data were available from 713 patients with SSc. Digital pitting was present in approximately half of these patients (n=362, 51%). Table 1 summarises their characteristics.

Table 1. Descriptive statistics of patients with and without digital pitting

	Digital Pitting (n=362)	No Digital Pitting (n=351)
Age, mean (SD)	67.1 (14.5)	66.5 (13.6)
Females, n (%)	287 (79.3)	301 (85.8)
Age at Raynaud's onset/ Age at SSc onset, median (IQR)	39.3 (25.2, 49.3)/ 41.4 (26.2, 52.5)	47.5 (35.3, 57.3)/ 52.5 (41.9, 60.4)
Limited/ Diffuse subtype, n (%)	276/85 (76.5/ 23.6)	257/93 (73.6/ 26.6)
History of IV vasodilators, n (%)	167 (46.4)	52 (14.9)
History of amputations, n (%)	52 (14.4)	5 (1.4)
Debridements, n (%)	72 (20.2)	18 (5.1)
Autoantibody status, n (%):	13 (6.2) ¹ / 61 (17.1) ³ / 152 (42.6) ⁵	34 (14.0) ² / 41 (12.0) ⁴ / 276 (79.3) ⁶
Anti-RNA polymerase/ Anti-Scl70/ Anti-centromere/ Anti-RNP	15 (4.3) ⁷	120 (35.4) ⁸ / 22 (6.5) ⁹

Denominator populations ¹210, ²246, ³357, ⁴345, ⁵357, ⁶341, ⁷352, ⁸341

From the univariable analysis (Table 2), gender (female, $p=0.02$), age at Raynaud's onset ($p<0.001$), age at SSc onset ($p<0.001$), IV vasodilators ($p<0.001$), amputations ($p<0.001$), debridements ($p<0.001$), anti-RNA polymerase ($p=0.01$), anti-Scl70 ($p=0.05$) and anti-centromere ($p=0.05$) were found to be significantly associated (anti-RNA polymerase negatively ($p=0.20$)) with digital pitting ($p\leq 0.05$). Further analysis adjusting the p value for multiple testing (Bonferroni adjustment, $p\leq 0.0036$) found age at Raynaud's onset, age at SSc onset, history of IV vasodilators, amputations and debridements to be significantly associated with digital pitting.

Table 2. Univariable logistic regression

	Odds Ratio	95% Confidence Interval	P value ¹	Number of patients in each analysis
Age	1.000	0.998 to 1.003	0.62	713
Female	1.116	1.015 to 1.231	0.02	713
Age at Raynaud's onset	1.008	1.007 to 1.010	<0.001	713
Age at SSc onset	1.004	1.003 to 1.005	<0.001	713
Limited/ Diffuse subtype	1.039/ 1.041	1.049 to 1.127/ 1.046 to 1.139	0.39/ 0.35	710/ 711
History of IV vasodilators	1.448	1.336 to 1.553	<0.001	710
History of amputations	1.553	1.363 to 1.768	<0.001	713
Debridements	1.405	1.259 to 1.568	<0.001	707
Autoantibody status:	1.223/ 1.111/ 1.052 to 1.419/ 1.000 to 0.01/ 0.05/ 456/ 702/ 698/			
Anti-RNA polymerase/ Anti-Scl70/ Anti-centromere/ Anti-RNP	1.081/ 1.114	1.234/ 1.002 to 1.162/ 1.310 to 1.786	0.05/ 0.20	693

¹ $p\leq 0.05$