Table 1. Time to diagnosis from Raynaud's and first non-Raynaud's symptom onset stratified clinical and demographic parametres

	Time to diagnosis from RF median-months	p	Time to diagnosis from first non-RF median-months	p
Sex				
Female	19 (4-299)	>0,05	11 (0-397)	>0,05
Male	39 (1-588)		15 (0-277)	
Education situation				
Less than highschool	44(0-588)	>0,05	11(0-397)	>0,05
Highschool and more	20(0-288)		14(0-297)	
Scl subtype				
Diffuse	23 (1-299)	***0.013****	11(0-297)	>0,05
Limited	45 (1-588)		11.5(0-397)	
Referred by healthcare practitioner				
Positive	24 (1-349)	>0,05	11(0-191)	>0,05
Negative	42 (1-588)		11(0-397)	
Initial symptom				
RF	49 (0-588)	***0.001***		
RF-non-RF together	21 (0-470)			

RF-non-RF together: less than one year between onset of RF and first non-RF symptom

Conclusions: Time to diagnosis from onset of RF is significantly shorter in diffuse SSc. Although mRS is typically higher in diffuse SSc, no correlation has been found between mRS and time to diagnosis. Therefore, duration of diagnosis may be influenced by internal organ involvement and other complications rather than skin changes due to lack of awareness of physicians about SSc related early skin changes. Referral to a proper physician by a familier healthcare practitioner decreased the number of physicians attending till the diagnosis, although it did not affect the time to diagnosis.

Disclosure of Interest: None declared

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AB0659 ASSOCIATION BETWEEN -12518A/G GENE POLYMORPHISM ENCODING MONOCYTE CHEMOATTRACTANT PROTEIN 1 (MCP-1) AND SERUM LEVEL OF C-REACTIVE PROTEIN IN DIFFERENT CLINICAL AND SEROLOGICAL PHENOTYPES OF SYSTEMIC SCLEROSIS IN THE RUSSIAN COHORT OF PATIENTS

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Background: Immune system activation with associated up-regulation in the production of extra-cellular matrix proteins by fibroblasts are known specific features in the pathogenesis of systemic scleroderma (SSc). Most recent data indicate that MCP-1 and MCP-3 chemokines from the family of monocyte chemoattractant proteins are also involved into SSc pathogenetic process. C-reactive protein (CRP) is known as the marker of acute-phase inflammation. The association between increased CRP levels and SSc clinical and serological parameters has been reported recently.

Objectives: To study the association between -2518 A/G gene polymorphism, encoding *MCP-1*, and CRP levels in different clinical SSc phenotypes in the Russian cohort of pts.

Methods: PCR-RFLP method was used to identify *MCP-1* genotype in 81 SSc pts aged 49,4±12,6 years, with mean SSc duration 11,1±9,0 years. CRP concentrations were measured with highly sensitive immunoturbidimertry method.

Results: CRP levels were correlated with *MCP-1* genotypes in pts with limited (lcSSc) and diffuse (dcSSc) phenotypes, with interstitial lung disease (ILD+), with SSc duration >3 years, with increased CRP level (>5 mg/L), with positive antibody titers to DNA topoisomerase I (ATA+) and antibody to centromeres (ACA+). A total cohort analysis showed that carriers of -2518AA genotype had higher mean CRP level versus G allele carriers (12,6±7,5 mg/L vs 4,9±4,8 mg/L, respectively, p=0,009), although similar trend was found in dcSSc phenotype (16,4±19,5 mg/L vs 6,1±4,4 mg/L, respectively, p=0,040). In pts with -2518AA genotype and SSc duration >3 years mean CRP level was significantly higher than in G allele carriers (1,1±16,7 mg/L vs 4,5±4,4 mg/L, respectively, p=0,025). In (ILD+) and (ATA+) subgroup pts with -2518AA genotype demonstrated higher mean CRP levels as compared to G allele carriers (12,4±15,6 mg/L vs 5,5±5,1 mg/L, respectively, p=0,018; and 17,6±20,9 mg/L vs 5,5±5,5 mg/L, respectively, p=0,010). CRP levels (>5 mg/L) were found in 31 (38%) pts and were significantly different between AA genotype carriers and G allele carriers (27,4±19,2 mg/L vs 10,4±3,8 mg/L, respectively, p=0,003). No associations between genetic variations in the *MCP-1* gene and CRP levels in lcSSc phenotype, SSc duration <3 years, CRP levels <5mg/L and (ACA+) pts were established.

Conclusions: Our data demonstrate that -2518A/G *MCP-1* gene polymorphism is closely associated with CRP levels, thus, it can be considered as a new marker, reflecting the severity of the disease and unfavorable SSc prognosis.

Disclosure of Interest: None declared

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AB0660 CHARACTERISTICS OF AL-I LIGHT-CHAIN AND AMYLOID A DEPOSITION IN PROGRESSIVE SYSTEMIC SCLEROSIS – A COMPARATIVE POSTMORTEM CLINICOPATHOLOGIC STUDY OF 12 PATIENTS

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Background: Different types of amyloid deposits may be present in systemic sclerosis (SSc), as consequences (complications) of basic or associated diseases.

Objectives: The aim of this study was to determine the type, prevalence and extent of amyloid deposits on different tissue structures in various organs in SSc.

Methods: We studied histopathologically 12 SSc patients (females 11, average age: 54.82 years, range 66–32, onset of SSc: 48.86, average disease duration: 6.43 years; one male, age 65.0 years at death, onset of SSc and average disease duration not known, who died at the National Institute of Rheumatology. SSc was diagnosed clinically according to the criteria of the ACR [1]. In 1 (8.0% of 12) 67 year old female patient (onset of SSc: 66 years, disease duration: 1 year) SSc was accompanied by B-cell lymphoma and complicated by systemic AL-l light-chain amyloidosis. In 1 (8.0% of 12) 53 year old female patient (onset of SSc: 41 years, disease duration: 12 years) SSc was complicated by systemic amyloid A (aA) deposition.

Amyloid deposits on different tissue structures [arteriole, small artery, medium size artery, venule, small vein, medium size vein, interstitial collagen fiber, reticulin fiber (collagen IV), and nerve] of 6 organs [heart, lungs, kidney, gastrointestinal tract, skin and brain] were determined histologically. The extent of amyloid deposition was evaluated by semi-quantitative, visual estimation on a 0 to 3 plus scale, based on the number of involved tissue structures per light microscopic field [2].

The prevalence and extent of amyloid-l light-chain and amyloid A deposits on different tissue structures were compared by Student (Welch) t-probe.

Results: The involvement of different tissue structures (prevalence in %) and the average extent of AL-l light-chain and amyloid A deposits (absolute value) are summarized in Table 1.

Table 1

Tissue structures	SSc-l Prevalence in %	SSc-aA Prevalence in %	p<	SSc-l Average extent	SSc-aA Average extent	p<
Arteriole	83,33	66,67	0,2749	2,00	1,33	0,1709
Small artery	83,33	66,67	0,1709	2,17	1,00	0,0414
Medium size artery	83,33	66,67	0,1520	2,33	0,67	0,0090
Interstitial collagen	66,67	50,00	0,2998	0,83	0,83	0,5000
Medium size vein	50,00	50,00	0,2735	1,17	0,50	0,1476
Small vein	50,00	50,00	0,5000	0,58	0,50	0,4088
Venule	33,33	33,33	0,5000	0,33	0,33	0,5000
Collagen IV	33,33	16,67	0,2749	0,50	0,33	0,3671
Nerve	0,00	0,00		0,00	0,00	
Average/Structure	53,70	44,44	0,232	1,10	0,61	0,075

Conclusions: In SSc patients the prevalence and extent of l light-chain and amyloid A deposits on different tissue structures changed parallel.

The higher prevalence and extent of l light-chain deposits in contrast to amyloid A may be explained with qualitative differences of l light-chain and amyloid A; l light-chain seems to have greater affinity for tissues than amyloid A protein.

Infiltration of the vessel walls – regarding the amount of l light-chain and amyloid A deposits in arterioles and arteries in contrast to the veins – showed a converse tendency in SSc patients with AL-l or AAa. This may be related to sluggish blood flow or stasis (backward congestion and accumulation of circulating precursors) in both diseases.

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AB0661 ORAL HEALTH-RELATED QUALITY OF LIFE MEASURED WITH OHIP 49 HIGHLY CORRELATES WITH DISEASE ACTIVITY AND SEVERITY IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is associated with decreased saliva production and interincisal distance, more missing teeth, and periodontal disease. Orofacial manifestations of SSc contribute greatly to overall disease burden and still are regularly overlooked and under-treated. Previous studies did not confirm correlation between disease severity and oral health-related quality of life in SSc patients.

Objectives: The aim of this study was to determine possible correlation of the SSc clinical parameters with oral health-related quality of life measured with the Oral Health Impact Profile 49 (OHIP 49).
Methods: Subjects were recruited from the Center of excellency for systemic sclerosis in Croatia cohort. Detailed dental by the same dentist and clinical examinations were performed according to standardized protocols. Associations between oral health-related quality of life and disease charachteristics were examined. We evaluated the disease severity using clinical and laboratory parameters according to the Medsger Severity Scale. The level of SSc activity was evaluated according to Valentini activity score. Oral quality of life was measured using the OHIP 49, which consists of 49 questions on the frequency of adverse oral conditions such as toothache, mouth pain, difficulty chewing or pronouncing words and discomfort related to appearance (higher scores indicating worse oral health-related quality of life). The study was approved by the University Hospital Split Ethics Committee.
Results: Thirty-one SSc patients (29 women and 2 men, mean age 56.45±13.60 years, median disease duration 7 years with minimum–maximum range 1–28 years) were consecutively enrolled for this study between January 2014 and December 2015. All patients fulfilled the ACR criteria for the diagnosis of SSc. The distinction between limited cutaneous SSc (lcSSc) and diffuse cutaneous SSc (dcSSc) was made according to the Leroy et al. criteria (28 dcSSc, 3 lcSSc). OHIP 49 scores highly positively correlated with disease activity (p=0.005, r=0.4872, Spearman's rank coefficient) and severity (p=0.016, r=0.4303, Spearman's rank coefficient). Furthermore, oral health-related quality of life positively correlated with the skin involvement evaluated by modified Rodnan skin score (p=0.003, r=0. 5207, Spearman's rank coefficient). Impaired quality of oral health positively correlated with the severity of general involvement, skin, gastrointestinal and joint/tendon involvement (p=0.003, r=0.506 for general involvement, p=0.003, r=0.511; $p<0.001$, r=0.591 and p=0.02, r=0.391 for skin, gastrointestinal and joint/tendon involvement, respectively, Spearman's rank coefficient). OHIP 49 score was highly variable between anti-topoisomerase I antibodies positive or negative SSc patients ($p<0.001$, Fisher's exact test).
Conclusions: Contrary to previous studies in our study disease severity and activity were related to OHIP 49 scores. Our data suggest that OHIP scores correlate with severity of general involvement, skin, gastrointestinal, and joint/tendon involvement in SSc patients. Disease subset and autoantibodies profil could play a role in the oral manifestation of SSc. Better collaboration between rheumatologists and the dental team is required to improve access to dental care and oral health outcomes for SSc patients.
Disclosure of Interest: None declared
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AB0662 SYSTEMIC SCLEROSIS (SSC) COHORT IN ABU DHABI: FOCUS ON DIGITAL ULCERS (DU)

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Background: DU are a significant burden for SSc patients affecting approximately 40–50% of patients (1). No data are available on the frequency of DU on SSC patients living in the United Arab Emirates.
Objectives: To identify the frequency of DU in a cohort of SSC in Abu Dhabi which is the capital city of the United Arab Emirates (UAE).
Methods: SSC patients identified according to ACR EULAR criteria through the hospital electronic medical records system, which was implemented from January 2011 for medical documentation in all public hospitals in Abu- Dhabi and districts. Using the International statistical classification of diseases and related health problems, usually called by the short form name international classification of Diseases (ICD), version 9 (710.1) from 1st of January 2011 to 31st of December 2016. Retrospective review of electronic medical records and paper case notes was performed on patients who presented during this period. The frequency of DU was searched. DU were defined (1) and categorised (2) (no digital ulcers, episodic (rarely recurrent), recurrent (frequently recurrent) and chronic ($\geq$1 DU at every follow up). Results and the incidence of gangrene were compared to data from DUO registry (3).
Results: 47 SSc patients (Male to female ratio 12 (25.5%) to 35 (74.5%) (Duration of disease from 2 to 24 years with peak age for ulcers between 40–50 years) were identified. No ulcers were detected in 34 patients (72.3%) while DU were found in 13 patients (27.7%): they were episodic in 9 patients (19%) and chronic in 4 patients (8.5%) of the total patients. No recurrent ulcers were found. Only one patient (2%) evolved to gangrene (she was a smoker). The other 12 patients with DU were non-smokers and three of them were under 16 years of age.
Conclusions: In Abu Dhabi SSc population, the incidence of DU is 27.7%. This differs from the incidence in the western literature where it peaks from 40 to 50% (3). Our patients had episodic (19%) and chronic DU (8.5%) of the total patients. The incidence of gangrene in our cohort was lower when compared to the data reported in the DUO registry (20%) (2). Although patients live in a very warm climate, DU are still experienced in SSc in Abu Dhabi. Likely, the chronic exposure to air conditioning may explain this paradox.
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AB0663 THE RELATIONSHIP OF SERUM LEVEL OF ANTICCP ANTIBODY WITH SEVERITY OF SKIN INVOLVEMENT IN SYSTEMIC SCLEROSIS PATIENTS REFERRED TO TOHID HOSPITAL

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Background: Systemic sclerosis is a chronic systemic disease of unknown etiology. There are a number of autoantibodies that correlated with disease severity and severity of skin involvement (1). Various studies have shown that AntiCCP Ab can also be seen in other autoimmune diseases such as systemic sclerosis (2–4). Results Polimen et al (4) and Morita et al (3) studies showed no relationship was found between AntiCCP Ab with skin Rodnon score.
Objectives: This study aimed to determine the relationship between serum levels of AntiCCP Ab with severity of skin involvement in systemic sclerosis patients referred to Tohid Hospital.
Methods: This study is a cross - sectional study, in the study population included all patients diagnosed with systemic sclerosis according to ACR 1980 criteria and subtypes according to Le Roy, that between 2013 and 2014 referred to Tohid hospital. We used from MRSS index to determine the extent of skin involvement. Sampling was available and we used to determine this association from chi-square statistics and to determine the amount of the connection from Phi statistics. Serum antibodies directed against CCP were assessed by ELISA.
Results: The results showed that the patients who were evaluated at the end of study were 50 patients, of whom 48 were women (96%) and (2%) 2 were male. The mean age of subjects is 38±10 years. The subtype of systemic sclerosis in 15 (%30 percent) of the patients were diffuse systemic sclerosis and 35 patients (%70 percent) have limited systemic sclerosis. Serum Anti CCP Abntibody was positive in 10% (5 patients). Also, there is no statistically significant relationship between the severity of skin involvement with Anticcp Ab (p=0.164).Severity of skin involvement in both groups was more severe form, and there is no statistically significant relationship between the severity of skin involvement and subtypes of systemic sclerosis (p=0.233). Also we didn't find no association between levels AntiCCP and joint symptoms and other symptoms.
Conclusions: In this study wasn't observed a significant correlation between serum levels of Anti CCP Antibody with severity of skin involvement in patients with systemic sclerosis (p=0.164),also serum level of AntiCCP Antibody couldn't as a predict factor to determine the severity of the disease. It seems that the relationship between other symptoms of systemic sclerosis and serum level of AntiCCP Antibody may be result from overlapping with RA.
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AB0664 LEVEL OF SATISFACTION IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES

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Background: Idiopathic Inflammatory Myopathies (IIM) are the group of rare diseases that carry a significant impact on patient's quality of life, influenced by the level of patient's satisfaction regarding medical services.
Objectives: To assess the patient's satisfaction and quality of life.
Methods: A cross-sectional study was performed from December 2015 to December 2016. There were included consecutive patients that fulfilled the Bohan and Peter[1] criteria for IIM. The collected information was about demographic data, clinical and laboratory findings. The patient's satisfaction was assessed by self-administered Patient Satisfaction Questionnaire (PSQ III)[2], which is a 50-item tool, covering 7 domains: general satisfaction, technical quality,