

the disease were noted. Also the social difficulties due to fingerprint abnormalities were noted. Healthy controls with no RP were included for comparison.

Results: 40 consecutive patients with RP attending Rheumatology outpatient services of our institute were screened for FP abnormalities. 29 with SSc [20-DcSSc, 9-LcSSc], 8 with overlap syndromes and 1 each of SLE, Undifferentiated Vasculitis and Undifferentiated CTD. It was noted prior to screening that 19 patients experienced some difficulty in the past with biometric recognition of their FPs at various times. On screening with biometric scanner, 15 of 40 [37.5%] had FP abnormalities in the form of non recognition of at least one finger with a median of 2 [range 1–6 fingers]. Of these 15, seven had DcSSc, six had LcSSc and two with overlap syndromes. The mean NFIQ score of these 15 patients was 4.5 [poor] and the mean NFIQ scores in SSc was 3.8. Eleven [27.5%] patients could not get government Identity cards based of FP scanning, four could not avail various government benefit schemes which needed their fingerprints as identity. Sixteen [40%] had history of digital vasculopathy in the form of digital pits, digital ischemia or ulcers. PAH was found in one and eight had Interstitial lung disease. Among the 10 controls all FPs were recognized and the mean NFIQ score was 2.2 indicating a better quality of FPs.

Conclusions: Fingerprint abnormalities occur frequently in patients with systemic sclerosis causing social disabilities in few. The quality of FPs in SSc patients is poor. Raynaud's phenomenon and vasculopathy are frequently associated. Documentation of this abnormality should allow the use of other biometric tools for personal identification.

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AB0645 LYMPHOCYTE SUBSETS T, B AND NK CELLS IN SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a rare multisystem disease with underlying immune mechanisms, whose pathogenesis remains unclear. Few previous reports have evaluated lymphocyte subpopulations in SSc and your results are conflicting.

Objectives: The present study aimed to analyze the lymphocyte subsets in SSc patients in comparison to healthy individuals.

Methods: Peripheral blood (PB) samples to analyze lymphocyte subsets were obtained from a non-random convenience sample of 20 SSc patients. Twenty healthy individuals recruited from the blood bank were used as sex and age-matched controls. Blood samples were analyzed by flow cytometry for total T cells, CD4+ and CD8+ T cells subsets, CD19+ B cells and total NK cells. Statistical analyses were performed using the IBM Statistical Package for Social Sciences (SPSS 18.0). Data are expressed as mean \pm SD and median and range. Non-parametric Mann-Whitney U test was used for analyses of the flow cytometry. A probability $p < 0.05$ was considered statistically significant.

Results: The mean (SD) age of SSc patients was 57.9 (14.2) years, 95% were female and 31.6% presented diffuse cutaneous SSc (dcSSc). Patients presented a lower mean total lymphocyte count compared to healthy controls (23.7% vs. 29.6%, $p = 0.026$) (Table 1.). No statistically significant differences were found in the percentages or the absolute numbers of T, B or NK cells.

		Patients	Controls	p^a
Leukocytes		8.02 \pm 1.51	6.79 \pm 3.39	0.108
Lymphocytes	%	23.53 \pm 14.06	29.97 \pm 15.08	0.026*
CD45	Absolute	1.70 \pm 1.59	2.06 \pm 0.03	0.512
	%	99.79 \pm 0.11	99.71 \pm 0.11	0.165
CD3	%	71.20 \pm 3.57	75.11 \pm 1.79	0.398
	Absolute	1.35 \pm 1.20	1.60 \pm 0.07	0.478
CD4	%	48.59 \pm 4.09	47.76 \pm 8.78	0.289
	Absolute	0.85 \pm 0.94	0.99 \pm 0.12	0.620
CD8	%	25.10 \pm 0.04	28.38 \pm 1.19	0.277
	Absolute	0.48 \pm 0.29	0.51 \pm 0.03	0.301
CD19	%	8.89 \pm 11.57	10.44 \pm 2.07	0.478
	Absolute	0.20 \pm 0.02	0.17 \pm 0.03	0.512
NK	%	6.73 \pm 8.39	6.71 \pm 5.40	0.883
	Absolute	0.14 \pm 0.35	0.14 \pm 0.09	0.947
Ratio	T/B	8.54 \pm 4.11	7.31 \pm 2.3	0.383
	CD4/CD8	1.93 \pm 0.23	1.73 \pm 0.33	0.157

Conclusions: Our data support previous reports indicating that depletion of lymphocyte in the PB of SSc patients. However, we found no significant difference in relation to lymphocyte subtypes, which differs from the literature data.

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Absolute count of T and B lymphocyte subsets is decreased in systemic sclerosis. Eur J Med Res 2010; 15:44–46.

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AB0646 DETERMINANTS OF QUALITY OF LIFE IN SYSTEMIC SCLEROSIS AND PATIENT'S PERCEPTION OF THEIR ILLNESS

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Background: Systemic sclerosis (SSc) is a chronic multi-system autoimmune disease associated with disability and reduced quality of life.

Objectives: The purpose of this study was to assess health-related quality of life and disease perception in a group of SSc patients.

Methods: We performed a case-control study on 50 SSc patients from EUSTAR cohort 096. Socio-demographic data, disease characteristics and self-assessment questionnaires: Health assessment questionnaire (HAQ), EuroQol-5D (EQ5D) and the Brief Illness Perception Questionnaire were collected.

Results: The group included 41 females, 31 limited SSc subsets.

Medium HAQ value was 0.9 (0.6). Patients with higher Rodnan score ($p = 0.002$), synovitis ($p = 0.02$), late capillaroscopic pattern ($p = 0.02$), muscle weakness ($p = 0.001$), gastrointestinal involvement ($p = 0.01$) and those on immunosuppressants ($p = 0.02$) have a poor life quality.

According to EQ-5D, the quality of life was related to specific organ involvement. 48% of the patients had some mobility problems, 6% were confined to bed; mobility was influenced by lung involvement ($p = 0.008$), digital ulcers ($p = 0.03$) and Medsger score ($p = 0.01$). 48% of the patients had some self-care problems and 8% were not able to wash/dry themselves; self-care was influenced by the Rodnan score ($p = 0.02$), diffuse subset ($p = 0.02$), muscle weakness ($p = 0.03$) and gastrointestinal involvement ($p = 0.021$). 64% of the patients had some problems in performing usual activities and 16% were not able to perform them; the performance was influenced by disease subset ($p = 0.01$), Medsger score ($p = 0.02$), cardiac involvement ($p = 0.02$) and the use of immunosuppressants ($p = 0.01$). 52% of the patients had some and 38% had extreme pain/discomfort; 66% of the patients were moderately and 20% were extremely anxious/depressed. Both were related to digital ulcers ($p = 0.01$, respectively $p = 0.045$).

The illness had a great impact on patients life 7.3 (2.5)/10. The main determinant was pulmonary fibrosis ($p = 0.04$). The patients consider that their disease will continue for quite a long time 8.7 (2.6)/10. Most of the patients do not feel to have a good control on their disease 6.3 (3.3)/10 and unfortunately they do not think that the treatment is very helpful 7.9 (2.7)/10. The intensity of the symptoms is quite severe 7.5 (2.7)/10, related to digital ulcers ($p = 0.04$) and gastrointestinal involvement ($p = 0.02$). Patients are very concerned about their disease 9.1 (2.3)/10, most of them being emotionally affected 7.6 (2.6).

Conclusions: This study confirms the presence and magnitude of impaired life quality in patients with SSc with impact on mobility, self-care, usual activities. The major determinants were the extend of skin involvement, musculoarticular, gastrointestinal involvement and digital ulcers. Often patients are anxious/depressed, had a high pain intensity and the perception of this illness is pessimistic.

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AB0647 DIAGNOSIS OF SYSTEMIC SCLEROSIS – “A TANGLED STORY”

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Background: Proper diagnosis of scleroderma is often long and difficult, since it is such a rare disease, and one which few doctors or patients are familiar with.

Objectives: To establish the interval between the symptoms' onset of systemic sclerosis (SSc) and what type of investigations are performed until the patients reach the final diagnosis of a rheumatologist

Methods: This is a cross-sectional study that included randomly selected patients with a diagnosis of SSc which were evaluated based on a questionnaire about symptoms at onset, specific consults and investigations. Descriptive statistics were used.

Results: The study group included 47 patients, of which only 5 were males and 17 from rural areas. The medium age was 53 (14.4) years.

First symptom of onset was Raynaud phenomena 91.3% of the cases, followed by skin changes (56.5%), puffy fingers (52.2%), gastrointestinal and musculoarticular symptoms (23.9% each).

The medium duration between the first symptom and a medical consult was 6 (63.5) months. The first medical consult was done by a internal medicine specialist -38.3%, by a rheumatologist-29.8%, a gastroenterologist-12.8%, a dermatologist-8.5%, a nephrologist and a pneumologist-4.2% and neurologist 2.1%

The first suspected diagnosis was SSc in 14 cases, Raynaud syndrome in 8, connective tissue disease in 5, rheumatoid arthritis in 4, cancer, autoimmune hepatitis, idiopathic pulmonary fibrosis each in 2 cases and none in 10 cases.

The medium number of consults until final diagnosis was 3.4 (1.7). The medium duration from the first symptom until the correct diagnosis was 39.2 (74) months. The first investigations recommended were blood tests in almost all of the patients (95.7%), but only a third of them included specific scleroderma autoantibodies. Capillaroscopy was performed as an initial diagnostic test in only 6 patients (12.8%). The mean interval from disease onset until the patient was referred to the first capillaroscopy was 13.5 (28.8) months, to specific autoantibodies was 40.17 (61.3) month, to echocardiography was 36.38 (54) months, to lung function tests and lung CT – 41.76 (65.8) months.

There were no significant statistical differences between patients coming from rural environment and those coming from urban environment. The only significant statistical difference between diffuse and limited subset was the time the patient was referred to echocardiography (19.8 (47.6) months for the diffuse subset, 66.8 (94.6) months for the limited subset, $p=0.04$).

The only statistical difference between males and females was related to the interval that capillaroscopy was performed (14 (20.5) months in females, 4.8 (5) months in males, $p=0.02$).

Conclusions: Scleroderma is a less well-known disease. This lack of awareness contributes to delayed diagnosis and delayed onset of therapy. Often such diagnostic uncertainty and frustration takes a huge toll on the psychological well-being of these patients, who describe their journey to diagnosis as being one of the most difficult part of their illness. One of our missions as rheumatologist is to increase recognition of this disorder.

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AB0648 DISEASE-RELATED AUTOANTIBODY PROFILE IN SYSTEMIC SCLEROSIS IN GREECE

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Background: Autoantibodies (autoAbs) help in diagnosis and predicting clinical phenotypes in systemic sclerosis (SSc).

Objectives: To determine the clinical utility of 13 SSc-related autoAbs in SSc patients in Greece.

Methods: 131 consecutive patients with SSc (111 female, mean age 58.1±14 years; 49 with diffused cutaneous SSc [dcSSc] and 82 with limited cutaneous SSc [lcSSc]) were analyzed by a multiplex line immunoassay (Euroimmun) for autoAbs against 13 SSc-related antigens. Twenty two patients with primary Raynaud phenomenon (RP), and 22 healthy controls were also analyzed

Results: ANA by indirect immunofluorescence was present in 128 (97.7%) patients with SSc. Excluding anti-Ro52, 113 (89.3%) SSc patients were positive for at least one autoAb: anti-Topo I abs in 54 (41.2%), anti-CENPA in 37 (28.2%), all reactive with CENPB), anti-RNA polymerase III (RP11) in 19 (14.5%), anti-RNA polymerase III (RP155) in 13 (9.9%), anti-fibrillarin in 4 (3.1%), anti-Ku in 6 (4.6%), anti-NOR90 in 8 (6.1%), anti-PM/Scl100 in 2 (1.5%), and anti-PM/Scl75 in 4 (3.1%). There was no immunoreactivity for Th/To or PDGFR. Overall, 102 (77.9%) SSc patients had autoAbs against Topo I, CENPA or CENPB, RP11 or RP155. Anti-Topo I abs were strongly associated with dcSSc, interstitial lung disease (ILD) ($p<0.001$), and pulmonary arterial hypertension (PAH) ($p=0.019$). Anti-CENB abs were associated with lcSSc, and negatively associated with ILD. Anti-RP11 and anti-NOR90 abs were associated with male gender, and anti-NOR90 associated with ILD. In PR individuals and healthy individuals anti-Ro52 immunoreactivity was found in 9.1% and 13.6%, respectively, but no other immunoreactivity was detected.

Conclusions: Anti-Scl70, anti-CENP and anti-RNA pol III are the most prevalent autoAbs in SSc. Anti-Topo I and anti-NOR90 abs are associated with ILD and/or PAH.

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AB0649 SYSTEMIC SCLEROSIS IN ARGENTINA: EVALUATION OF A SINGLE CENTER COHORT AND COMPARISON WITH INTERNATIONAL SERIES

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Background: Systemic sclerosis (SS) is an autoimmune disease with generalized vascular dysfunction and a myriad of clinical and laboratory manifestations. Spreading of thickening of the skin, serological characteristics and the pattern of compromise of the internal organs help to classify them as diffuse scleroderma, localized sclerosis and systemic sinus scleroderma. Reports of Latin American cohorts published to date are scarce.

Objectives: To describe the clinical and serological data for a single center cohort and to compare those with national and international cohorts.

Methods: Descriptive, observational, cross-sectional study. We analyzed our SS database. Patients were evaluated since 01/15 to 05/2016 and fulfilled SS classification criteria (ACR 1980/ACR-EULAR 2013). Patients were classified according to Le Roy criteria in Limited Systemic Sclerosis (ISS), Diffuse Systemic Sclerosis (dSS), Systemic Sclerosis sine scleroderma (SsnS) and pre-systemic sclerosis (pSS). Organic compromise was defined as follows: Gastrointestinal (GI): esophageal dysmotility by manometry or esophagitis by endoscopy; Lung: Pulmonary hypertension (PH): PSAP>25 mmHg by right heart catheterization; Interstitial Lung Disease (ILD): compatible HRCT or <70% predicted FVC or <80% predicted DLCO; Heart: Left ventricle dysfunction without PH or arterial hypertension, pericardial effusion; Digital ulcers and pitting scars; History of sclerodermic renal crisis (RSC); calcinosis. Anti-Scl-70 was determined by ELISA and anti-Centromere (ACA) by IIF.

Results: 123 patients were included. 74% with ISS, 24% dSS and 2% SsnS; age at diagnosis (years): 49.7 (18–79 DS: 12.27) and 48.7 (27–79 DS: 13.01) in ISS and dSS respectively. Raynaud's phenomenon previous to diagnosis (years) 7.7 (0–54 DS: 12.81) in ISS and 4.03 (0–22 DS: 5.5) in dSS. 49.5% ISS were ACA positive vs 17% in dSS as well also 41.4% had Scl-70 positive ($p<0.05$). Lung: 58.6% of dSS presented ILD and 3.4% RSC, according to published reports. No statistically significant differences were found about the presence of calcinosis, digital tip ulcers or PH between dSS and ISS. GI: was studied in only 39/123 patients with 84,6% affected among those.

Table 1. Comparison of main variables among different cohorts

	Diffuse				Limited systemic sclerosis					
	US, n		EUSTAR, n		Spain, n		Germany, n		Argentina*, n	
	119	128	2838	4481	243	568	484	674	29	91
Oesophageal dysmotility (%)	67	67	70	66	71	57	69	59	31.5	38.5
ILD (%)	63	37	52	11	70	39	56	21	59	22
PH (%)	2	31	6	ND	14	9	19	15	3.4	3.3
RSC (%)	17	2	4	1	8	1	16	9	3	0
Digital ulcers (%)	ND	ND	20	33	64	39	34	24	10.3	5.5
Calcinosis (%)	23	42	ND	ND	24	20	ND	ND	14	19

*HCJSM.

Conclusions: Our data showed similar results to the local and european cohorts. We found fewer calcinosis and digital tip ulcers in both ISS and dSS than international cohorts. A lower prevalence of PH was reported in ISS respect to other series, and it might be attributed to the method wch it was measured. (right heart catheterism).

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AB0650 ANTI-TOPOISOMERASE POSITIVE SYSTEMIC SCLEROSIS PROGNOSIS INFRAST?

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Background: Anti-topoisomerase antibodies (ATA) in systemic sclerosis (SSc) have been associated with poorer prognosis including diffuse skin involvement, pulmonary fibrosis, cardiac involvement and increased mortality. However, 30–60% of ATA-positive SSc patients demonstrate only limited skin involvement and some have only mild disease course. In SSc, optimal risk stratification is of utmost importance for tailored clinical management at the patient level.

Objectives: We aimed to determine the prevalence of mild disease among ATA positive patients and to investigate which readily available clinical parameters best identify patients with highest disease severity.

Methods: Clinical baseline data from SSc patients included in the Combined Care In Systemic Sclerosis (CCIS) cohort of the Leiden University Medical Center were extracted. Patients fulfilling ACR 2013 criteria and ATA positive were included. Descriptive statistics were used to summarize sociodemographic, clinical and serological features. Patients with grade 3 or 4 disease on any of the Medsger severity subscales were considered to be severely diseased. We compared presence of diffuse cutaneous involvement, Raynaud's, pitting scars, calcinosis, proximal muscle weakness, >10% weight loss, interstitial lung disease and pro-BNP between the severity groups and corrected for confounding by disease duration (time since non-Raynaud) by stratifying into quartiles.