SAT0291

PULMONARY INVOLVEMENT AND FUNCTIONAL LIMITATION IN SYSTEMIC SCLEROSIS

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Background: Pulmonary involvement is the main cause of death in Systemic Sclerosis patients (SS). However, there is little information whether its presence is capable of affecting the functional capacity of patients and if this influences the quality of life perceived by them.

Objectives: To determine whether the presence of pulmonary involvement in patients with SS (Interstitial Lung Disease and or Pulmonary Arterial Hypertension) is related to greater functional disability.

Methods: Observational and cross-sectional study, with a prospectively performed protocol, of patients diagnosed of SS according to ACR/EULAR 2013 criteria. Demographic, clinical, analytical, activity (EUSTAR index), severity (Medsker scale and modified Rodnan index), health perception (SF36) and disability (HAQ and Cochin test) variables were collected. Moreover, Videocapillaroscopy (VCL) and Respiratory Function Test were made. All the patients had pulmonary TCMD and echocardiography in order to describe pulmonary features.

Results: 42 patients were included (95.4% women), with an average age of 59.2 (S.D 12.9) years. The median of years from diagnosis was 4, and 6 from the first not Raynaud symptom. Out of them, 20 were SS limited, 20 patients SS diffuse; and 2 patients SS sine scleroderma. Pulmonary hypertension (HTP) was found in 11.9% of patients, as well as EPID in 33.3% (UIP 16.7%, NSIP 14.3%, bronchiolitis 2.3%). Analyzing the subgroup of patients with EPID, we can highlight higher score of HAQ, Cochin and activity index EUSTAR, with less influence in Rodnan index and without differences in SF36.

	ILD patients	Non-ILD patients	PAH patients	Non-PAH patients
HAQ (median)	0.75	0.44	1.25	0.5
EUSTAR activity index (median)	6	4	6.3	4
Cochin (median)	10.5	5	5	5
SF36 (median)	0.52	0.52	0.42	0.44
Rodnan (median)	3	2.5	4	3
Medsker Severity (median)	4	4	5	4
PAH (%)	7.1	14.2	20	35.1
Centromeric nuclear ANA pattern (%)	7	71	40	51.3
Homogeneous nuclear ANA pattern (%)	36	14	20	24.3
Anti-centromere (%)	7	71	40	51
Anti-Sc170 (%)	36	7	20	16
Anti-RNAPolymeraseIII (%)	14	7	20	8.1
Reduction in DLCO (%)	71	53	80	56.8
Reduction in FVC (%)	21	14	40	13.5
Sclerodermaform pattern en VCL (%)	78.6	85.7	100	81.1

Regarding the subset of patients with PAH, we found higher score on HAQ, EUSTAR activity index, index Rodnan, Medsker gravity scale and the SF36, than in non-PAH patients, with no differences in Cochin test.

Conclusion: We can highlight the presence of greater functional limitation that is especially reflected in rates of disability (HAQ and Cochin), as well as a increased activity of disease and a lower self-perception of health in patients with Systemic Sclerosis pulmonary involvement.

Disclosure of Interests: None declared

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SAT0292

SELF-ADMINISTERED VERSUS INTERVIEW BASED INTERNATIONAL CONSENSUS CRITERIA FOR THE DIAGNOSIS OF RAYNAUD'S PHENOMENON QUESTIONNAIRE – A SINGLE CENTRE EXPERIENCE

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Background: International Consensus Criteria for diagnosing Raynaud's Phenomenon (RP) have been proposed recently (1). These criteria are based on the three-step conditional approach (i.e. a screening question,

assessment of colour changes and an item of disease score calculation). It is unclear whether differences exist between the self-administered and interview-based way of questionnaire fulfillment.

Objectives: We aimed in our study to evaluate the two approaches in patients with suspected RP.

Methods: This cross-sectional study was conducted at our secondary/tertiary rheumatology centre between 1 October 2018 and 31 December 2018. Each patient referred for the video capillaroscopy was asked to complete the RP questionnaire on the day of the referral. The same questionnaire was applied as an interview by the rheumatologist before the video capillaroscopy. Differences in answers and level of individual agreement on individual three items, as well as the result of the questionnaire, between the two ways of questionnaire completion were assessed with Fisher exact test and Kappa coefficient, respectively.

Results: During the 3-month period we included 84 consecutive patients (88.1% female, median age (IQR) 49.5 (41.9–56.5) years). Results of answers to each item, final outcome of the questionnaire and the level of individual agreement for each item are presented in Table 1.

Table 1.. Answers to Raynaud's phenomenon International Consensus Criteria questionnaire according to mode of administration

	Self-administered Yes, N (%)	Interview based Yes, N (%)	p-value**	Measure of individual agreement– Kappa coefficient (95% CI)
Item 1. Are your fingers unusually sensitive to cold?	79 (94.0%)	78 (92.9%)	1	0.514 (0.101–0.927)
Item 2. Occurrence of biphasic colour change during the vasospastic episodes (white and blue)	40 (47.6%)	42 (50.0%)	0.877	0.762 (0.623–0.900)
Item 3. RP disease score	53(63.1%)	67 (79.8%)	0.026	0.549 (0.330–0.747)
Presence of RP according to RPICq	31 (36.9%)	36 (42.9%)	0.529	0.777 (0.640–0.915)

*RP – Raynaud's phenomenon; **RPICq** - International Consensus Criteria for the Diagnosis of Raynaud's Phenomenon questionnaire, ** Fisher-exact test

Conclusion: Overall, there was a substantial level of agreement between self-administered and interview-based application of International Consensus Criteria for the Diagnosis of Raynaud's Phenomenon questionnaire. However, we observed that the patients were inclined to score the severity of their RP lower than the physicians.

REFERENCE

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SAT0293

SPECIFIC ALTERATIONS OF NAILFOLD CAPILLARIES MIGHT ANTICIPATE THE APPEARANCE OF THE EARLY CAPILLAROSCOPIC SCLERODERMA-PATTERN IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Non-specific abnormalities (i.e. capillary enlargements) are usually found at the nailfold videocapillaroscopy (NVC) analysis in patients affected by primary Raynaud's phenomenon (PRP). We have previously demonstrated that capillary diameter is an independent predictor for development of systemic sclerosis (SSc) associated secondary Raynaud's Phenomenon (SRP), so that progression to SRP is unlikely for subjects affected by RP when average nailfold capillary diameter is under 30 µm [1]. However, until now these findings have not been clearly classified in a defined pattern able to really predict the evolution to a scleroderma-pattern.

Objectives: This pilot study aimed to identify in a cohort of SSc patients a "very early" NVC pattern at high risk of evolution in the already defined NVC scleroderma-patterns.

Methods: We selected the NVCs of 273 patients affected by SSc (according to 2013 ACR criteria) who presented one of the validated NVC scleroderma patterns (81 with "Early" pattern, 84 with "Active" pattern, 92 with "Late" pattern, 16 with "scleroderma-like" pattern) (2). Among the 273 SSc patient with an established NVC scleroderma-pattern, a number of 54 subjects who had previously NVC analyses performed before the development of the scleroderma-pattern were enrolled. Time of evolution was calculated, and a detailed pilot study of capillaroscopic characteristics was random executed on 10 of those patients. The analysis included the number and the limbs diameters (arterial, venous, and apical) of capillaries with a diameter over 30 µm, together with the total number of capillaries and microhemorrhages, in 16 images per subject.

Results: All the 54 patients (100%) showed enlarged capillaries with an average diameter over 30 µm in their previous NVC. At the follow up, thirty-one patients (57%) developed an "Early" scleroderma pattern in the following 3 years, 6 patients (11%) developed an "Active" pattern in 4 years, 3 patients (6%) evolved in "Late" NVC pattern in 5 years, whereas 14 patients (26%) developed a "scleroderma-like pattern" in 4 years. The average time of evolution in a scleroderma-pattern was 4 years. The detailed pilot morphological analysis conducted on 10 patients revealed an average total number of capillaries of 8.6/mm at last non-specific NVC analysis; among these 2.66 (31%) capillaries showed a diameter over 30 µm. The mean value diameter of the most dilated capillary was 35.74 µm (arterial limb 33.34 µm, apical limb 43.94 µm, venous limb 30 µm); the mean value for microhemorrhages was 0.6/mm. The mean number of capillaries reduced from 8.6±0.8 to 6.9±2.2/mm ($p=0.01$) during follow-up (4 years).

Conclusion: Present detailed pilot study demonstrates that SSc patients showed a significant increase of nailfold capillary diameter over 30 µm before development of a validated NVC scleroderma-pattern at follow up. Even if this is an independent predictor for development of SSc as previously demonstrated, the data could help in intercepting patients with RP at higher risk of evolution in a validated SSc NVC pattern.

REFERENCE

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SAT0294

ANALYSIS OF POLIAUTOIMMUNITY IN THE DIFFERENT SUBSETS OF SCLERODERMA

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Background: Coexistence of different connective tissue diseases based on their common autoimmune base is a feasible circumstance, which is known as poliautoimmunity (PAI).

Objectives: Evaluation of the occurrence of PAI in systemic sclerosis (SSc) according to subtypes in the patients included in the Spanish SSc Registry (RESCLE). Causes of death were also analyzed.

Methods: A nationwide, cross-sectional study was carried out. All participating centers had obtained local ethics committee approval.

Results: PAI was present in 46% out of 1911 patients, of whom, 33% had more than one association. Most of these patients were women

Table 1.. Prevalence of PAI according to subsets.

	Isolated SSc	PAI	P-VALUE
lcSSc	588 (57%)	554 (63%)	0.008
dcSSc	210 (20%)	188 (21%)	0.611
ssSSc	121 (12%)	85 (9.7%)	0.160
earlySSc	28 (2.7%)	16 (1.8%)	0.222
preSSc	85 (8.2%)	36 (4.1%)	<0.001

(93%, $p<0.001$), with no significant differences regarding first manifestation: Raynaud's phenomenon, puffy hands, arthralgia or skin sclerosis. Isolated SSc was more frequent in all subsets (table 1). Sjogren's syndrome was by far the most frequent associated disorder, with a RR of 1376 (table 2). Causes of death (SSc-related or not) did not significantly differ whether PAI was present or not.

Table 2.. Prevalence of the different associated autoimmune disorders.

	TOTAL	lcSSc	dcSSc	sineSSc	earlySSc	preSSc
Sjögren's syndrome	475 (25%)	317 (28%)	106 (27%)	34 (17%)	6 (14%)	12 (9.9%)
Thyroid autoimmune disorder	268 (14%)	173 (15%)	36 (9.0%)	35 (17%)	8 (18%)	16 (13%)
Primary biliary colangitis	88 (4.6%)	61 (5.3%)	5 (1.3%)	17 (8.3%)	0 (0.00%)	5 (4.1%)
Sclerosing colangitis	6 (0.31%)	4 (0.35%)	0 (0.00%)	0 (0.00%)	0 (0.00%)	2 (1.7%)
Autoimmune hepatitis	20 (1.0%)	17 (1.5%)	2 (0.50%)	0 (0.00%)	0 (0.00%)	1 (0.83%)
Systemic lupus erythematosus	11 (0.58%)	7 (0.61%)	1 (0.25%)	3 (1.5%)	0 (0.00%)	0 (0.00%)
Antiphospholipid syndrome	16 (0.84%)	8 (0.70%)	5 (1.3%)	2 (0.97%)	1 (2.3%)	0 (0.00%)
Inflammatory myopathy	99 (5.2%)	45 (3.9%)	47 (12%)	6 (2.9%)	1 (2.3%)	0 (0.00%)
Rheumatoid arthritis	6 (0.31%)	4 (0.35%)	2 (0.50%)	0 (0.00%)	0 (0.00%)	0 (0.00%)

Conclusion: A rather higher prevalence than reported was observed, although distribution of the associated disorders was similar. No remarkable differences were found regarding SSc subsets, either PAI prevalence, initial manifestations or causes of death.

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SAT0295

ABSTRACT WITHDRAWN

SAT0296

FAST TRACK ALGORITHM: HOW TO DIFFERENTIATE A SCLERODERMA PATTERN FROM A NON-SCLERODERMA PATTERN

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Background: The European League Against Rheumatism Study Group on Microcirculation in Rheumatic Diseases is a non-profit international network of expert centres. Its main research focus is to investigate the