Conclusion: Using a SLR and modified nominal technique, we have developed a preliminary pSScHI consensus-based definition and started a validation process for it to be used in clinical research and clinical practice.

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AB0557

CO-EXISTENCE OF SYSTEMIC SCLEROSIS HALLMARK AUTOANTIBODIES ASSOCIATES WITH DISTINCT CLINICAL PHENOTYPE

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Background: Systemic sclerosis (SSc) is typically manifests with distinct SSc-specific antibodies (SSc-Abs): anti-topoisomerase I (ATA), anti-centromere (ACA), anti-RNA polymerase III (ARA), anti-U3RNP (u3RNP), anti-U1RNP (U1RNP), anti-PmScl (PmScl), anti-Ku (Ku) and anti-Th/T0 (Th/T0), each being characterised by different clinical features and prognosis. The presence of >1 SSc-Abs is rare with minimum data about these patients' clinical phenotype.

Objectives: To describe and compare the clinical features of SSc patients with >1 SSc-Abs

Methods: The autoantibody profiles of 2799 SSc patients from February 2001 to June 2017 were retrospectively reviewed. Patients with >1 SSc-Abs were identified. Clinical features were collected and compared to historical cohorts of SSc patients with single SSc-Ab positivity. Patients were excluded if treated prior to their immunology test with rituximab, iv immunoglobulins or stem cell transplantation. Statistical analysis was performed using Fisher exact test.

Results: 72 patients (2.6%) with >1 SSc-Ab were identified. Full clinical data were available for 63 patients. 60 patients (2.1%) had double Ab positivity and 3 patients had triple Ab positivity (0.1%). 13 Ab combinations were present. U1RNP and ATA was the most frequent combination (35%), patients were significantly younger (51.38 years) than both U1RNP (58.64 years, $p=0.050$) and ATA (62.03 years, $p=0.002$) patients and more commonly of diffuse subset (dcSSc) ($p=0.001$ and $p=0.041$ respectively). Compared to ATA patients overlap features were more frequent (43% vs 15%, $p=0.004$) including inflammatory arthritis ($p=0.025$) and myositis ($p=0.013$) (Table 1). U1RNP and ACA had a significantly higher prevalence of pulmonary arterial hypertension compared to U1RNP (young) and ACA ($p=0.022$) patients, and compared to ACA patients they were younger (57.88 vs 68.75, $p=0.015$) with a higher incidence of myositis ($p=0.001$). U1RNP and ARA patients were more frequently dcSSc subtype compared to U1RNP patients (75% vs 21%, $p=0.040$). U1RNP and PmScl patients had a higher prevalence of myositis compared to U1RNP patients ($p=0.006$). ATA and ACA patients behaved similarly to ATA patients with a significantly higher prevalence of lung fibrosis ($p=0.006$) and myositis ($p=0.041$) compared to ACA. ACA and PmScl (7%) had higher prevalence of myositis compared to ACA patients ($p=0.04$).

Table 1. Frequency of clinical features in some of the double antibody group combinations, compared to our cohort of patients with only one of the SSc specific antibody. Significant p values (<0.05) highlighted in bold. ILD (interstitial lung disease), PAH (pulmonary arterial hypertension), SRC (scleroderma renal crisis).

Double antibody group	Comparator group	Clinical features											
		Arthritis		ILD		PAH		SRC		Cardiac		Myositis	
		n	p	n	p	n	p	n	p	n	p	n	p
U1RNP and ATA	U1RNP	6 (29%)		15 (68%)		1 (9%)		1 (1%)		0		4 (19%)	
	ATA	14 (18%)	1	38 (50%)	0.136	12 (16%)	0.287	1 (1%)	1	2 (3%)	1	12 (16%)	0.744
U1RNP and ACA	U1RNP	2 (10%)		2 (10%)		4 (20%)		0		0		1 (10%)	
	ACA	14 (18%)	1	38 (50%)	0.267	12 (16%)	0.039	1 (1%)	1	2 (3%)	1	12 (16%)	0.148
U1RNP and ARA	U1RNP	23 (30%)	0.087	43 (11%)	0.233	56 (15%)	0.022	6 (2%)	1	10 (3%)	1	3 (1%)	0.001
	ARA	0		2 (50%)		0		1 (25%)		1 (25%)		0	
U1RNP and PmScl	U1RNP	14 (18%)	0.311	38 (50%)	1	12 (16%)	1	1 (1%)	0.098	2 (3%)	0.144	12 (16%)	1
	PmScl	4 (1%)	1	64 (45%)	1	15 (10%)	1	34 (24%)	1	4 (3%)	0.131	5 (3%)	
ATA and ACA	ATA	1 (1%)		2 (66%)		12 (16%)	1	1 (1%)		2 (3%)		12 (16%)	0.006
	ACA	7 (13%)	1	27 (50%)	1	3 (6%)	1	3 (6%)	1	1 (2%)	1	21 (39%)	0.009
ACA and PmScl	ACA	28 (10%)	1	213 (79%)	1	12 (4%)	1	17 (6%)	1	20 (7%)	1	10 (4%)	0.353
	PmScl	23 (30%)	1	43 (11%)	0.006	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	0.041
ACA and U1RNP	ACA	1 (25%)		1 (25%)		0		0		0		1 (25%)	
	U1RNP	23 (30%)	0.227	43 (11%)	0.385	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	0.04
ACA and U1RNP	ACA	7 (13%)	0.457	22 (50%)	0.612	3 (6%)	1	3 (6%)	1	1 (2%)	1	21 (39%)	1
	U1RNP	0		1 (50%)		0		0		0		0	
ACA and U1RNP	ACA	23 (30%)	1	43 (11%)	0.216	56 (15%)	1	6 (2%)	1	10 (3%)	1	3 (1%)	1
	U1RNP	3 (6%)	1	12 (24%)	0.456	13 (26%)	1	5 (10%)	1	6 (12%)	1	9 (18%)	1

Conclusion: Coexistence of hallmark autoantibodies is exceedingly rare in SSc patients. When combined, both SSc-Abs have the potential to synergistically interact and modify the clinical phenotype.

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AB0558

RISK FACTORS FOR THE DEVELOPMENT OF BREAST CANCER IN SYSTEMIC SCLEROSIS

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Background: Prior literature shows a global increase of cancer risk among patients with systemic sclerosis (SSc). Although breast cancer (BC) is one of the most frequent malignancies in SSc patients, the characteristics of this neoplasm among SSc patients is not well established and it is uncertain whether SSc significantly increases the risk of this cancer.

Objectives: Describe the characteristics and risk factors for BC among Spanish patients with SSc.

Methods: Ambispective and multicenter study of patients with a diagnosis of SSc according to ACR/EULAR criteria and/or LeRoy classification included in the Spanish Scleroderma Registry (RESCLE) from 2006 to 2018. Characteristics of patients with BC were described and incidence was compared with that of the general population.

Results: Among 1930 patients with SSc, 206 (10.7%) had cancer. BC was the most frequent tumor location (47 patients of 206 with cancer [22.8%]), followed by lung cancer (29, 14.1%). The risk of BC was increased in patients with SSc compared to the general population (standardized incidence ratio [SIR] 1.31; 95% CI 1.10-1.54; $P = 0.003$).

The comparison of patients with BC and those without cancer showed that patients with BC had older age at diagnosis of SSc (50.9 vs 45.9 years, respectively; $P = 0.004$), were more frequently diagnosed of interstitial lung disease (ILD) (30/47 [63.8%] vs 694/1714 [40.5%]; $P = 0.002$) and pulmonary hypertension (12/24 [50.0%] vs 262/917 [28.6%]; $P = 0.037$), had higher frequency of puffy hands as the first manifestation of SSc (4/45 [8.9%] vs 38/1664 [2.3%]; $P = 0.023$) and had more frequently primary biliary colangitis (PBC) (7/46 [15.2%] vs 72/1708 [4.2%]; $P = 0.004$). Regarding autoimmunity profile, patients with SSc and BC had a significantly higher presence of anti-Ro (11/44 [25.0%] vs 214/1528 [14.0%]; $P = 0.049$) and anti-mitochondrial antibodies (7/28 [25.0%] vs 96/837 [11.5%]; $P = 0.039$). Multivariable regression analysis showed an independent association between the puffy hands (OR = 6.40; 95% CI 1.73-23.60; $P = 0.005$), diagnosis of PBC (OR = 5.70; 95% CI 2.16-15.07; $P = 0.001$), presence of ILD (OR = 3.29, 95% CI 1.69-6.39; $P < 0.001$) and the presence of the anti-Ro antibody (OR 2.14; 95% CI 1.01-4.56; $P = 0.048$) with the presence of BC.

Conclusion: BC risk was increased in patients with SSc. The development of ILD, PBC, the presence of anti Ro and puffy hands as the first clinical manifestation of SSc were identified as independent factors associated with the development of BC in our cohort.

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AB0559

ARRHYTHMIAS AND LEFT VENTRICULAR DYSFUNCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS.

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Background: Cardiac involvement in systemic sclerosis (SS) is common and one of the leading causes of mortality (up to 31%). Usually, it appears early and in a silent way. It can affect any cardiac structure and present with various manifestations. Cardiac disease worsens the prognosis and increases mortality, so it should be kept in mind in these patients.

Objectives: Study the prevalence of arrhythmias in patients with SS, not affected with pulmonary arterial hypertension (PAH) or other cardiac diseases, and establish its association with left ventricular (LV) dysfunction and heart dysautonomia (HD), as well as with other variables that could predict the development of this complication.

Methods: Prospective study of a cohort of patients with SS, excluding those with heart disease, PAH or cardiovascular risk factors. All underwent a clinical assessment, blood test with cardiac biomarkers, electrocardiogram (ECG), Holter 24h (HLT) and echocardiogram (TTE), interpreted by an expert cardiologist blind about the patients. Arrhythmias were classified as clinically significant arrhythmias (CSA) or clinically nonsignificant arrhythmias (CNSA) by ECG and HLT. LV diastolic dysfunction (LVDD) was defined as $E/e' > 8$, LV systolic dysfunction (LVSD) as a global longitudinal strain $< 20\%$ and HD as a SDNN < 100 ms. Demographic, clinical and biological data were collected. A follow-up was performed at 6.2 ± 0.9 years. Statistical analysis was performed using SPSS 23 IBM®.

Results: 36 patients were included: age 56.7 ± 12.3 years (y), male / female 35/1, disease duration 7 ± 4.1 y. 66% belonged to the limited SS subtype, 66.6% were anti-centromere+, 25% anti-topoisomerase, 2.7% anti-PM/Scl and 2.7% anti-RNA polymerase III+. Raynaud was present in a 100%, telangiectasia in 55.6% and interstitial lung disease in 36.1%. The modified Rodnan skin score (mRSS) was $\leq$ moderate (0-29 points) in 55.6%; 27.8% had presented digital ulcers that required prostaglandins.

27.8% had LVDD, 22% LVSD, 11.1% LVDD + SD and 16.7% HD. 50% (18/36) of patients had ECG alterations, of which 44% corresponded to CSA (Table 1) and, 55.6% (20/36) HLT alterations, of which 75% were CSA (Table 2). 3/36 patients had both HLT and ECG CSA. In 1 patient, impaired LV ejection fraction was detected; in none, valvular disease. 38.8% had elevated NT-proBNP and 13.9% troponin T (TnT). No correlation was found between any parameters and CSA.

A correlation was found between mRSS and DLCO ($p=0.002$), DLCO and digital ulcers ($p=0.001$), and mRSS and digital ulcers ($p=0.005$). A correlation was also found between elevated NT-proBNP and TnT ($p=0.006$) and between elevated NT-proBNP and LVDD ($p=0.049$).

At follow-up after 6.2 ± 0.9 y, 2 patients had died: 1 of neoplasia and 1 of severe biventricular dysfunction 5.2 y after the study.

Conclusion: Our data confirm a high prevalence of ventricular arrhythmias and left dysfunction in patients with SS, without heart disease, cardiorespiratory symptoms or HAP, being up to 75% of the arrhythmias CSA. The lack of correlation between CSA and LVSD or DD indicates that arrhythmias could be due, not only to a supposed structural alteration of the myocardium, but to a primary and early cardiac involvement in SS. In addition, the lack of correlation between CSA in ECG and HLT reinforces the importance of a complete cardiac evaluation in these patients to rule out silent cardiac involvement.

Table 1. ECG abnormalities. *CSA.

Alterations	# Patients
Major	Complete Left Bundle Branch Block (BBB)* 1
Minor	Abnormal QRS prolongation in precordial leads 1
	Nonspecific ST-T wave changes 3
	Incomplete Left BBB* 3
	Incomplete Right BBB* 4
Other	QT prolongation 11

Table 2. HLT alterations. *CSA.

Alterations	Subtype	# Patients
Supraventricular Extrasystoles	Uncommon	7
	Frequent*	3
	Mono/Polymorphic	0
Nonsustained Supraventricular Tachycardia*		7
Ventricular Extrasystoles	Benign	4
	No Benign*	4
	Doubles	1
	Two Morphologies	2
	Accelerated Idioventricular Rhythm	1
1st-degree Atrioventricular Block		1
Sinus Tachycardia		2

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