$r = -0.412$, respectively) and with hip fracture risk with and without FN-BMD ($r = -0.799$, $r = -0.412$, respectively). Major fracture risk with and without FN-BMD presented a moderate correlation with spine fractures ($r = 0.350$; $r = 0.397$, respectively).

No correlation was found between WHO threshold and spine fractures. No correlations were found between FN-BMD or fracture risk estimated by FRAX and disease manifestations, anti-Scl70 or ACA positivity, vitamin D insufficiency, smoking or GCT use.

Conclusion: In our cohort, low BMD was prevalent and had correlation with BMI. FRAX appears to be an useful instrument as it correlated with spine fractures, contrary to what was verified when we used the WHO threshold. Early monitoring of BMD and estimating fracture risk using FRAX appear to be useful tools for the prevention of fractures in this population.

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SAT0321

CURRENT PATIENT REPORTED OUTCOMES (PROS) POORLY REFLECT CHANGES IN LUNG FUNCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS

F. Danzo¹, K. Gjeshi¹, G. Abignano^{2,3,4}, A. M. Dean³, F. Masini¹, G. Cuomo¹, F. Del Galdo^{3,4}. ¹Università degli Studi della Campania, L. Vanvitelli, UOC Medicina Interna, Naples, Italy; ²Istituto Reumatologico Lucano (IReL), Azienda Ospedaliera Regionale San Carlo, Potenza, Italy; ³Leeds Institute of Rheumatic and Musculoskeletal Medicine, University of Leeds, Leeds, United Kingdom; ⁴NIHR Leeds Biomedical Research Centre, Leeds Teaching Hospital, Leeds, United Kingdom

Background: Lung involvement is very common in systemic sclerosis (SSc). Approximately one quarter of patients develops pulmonary problems within the first 3 years of diagnosis and still represents the leading cause of death in these patients. In a recent clinical trial, the reduction of FVC was not accompanied by a benefit with respect to health-related quality of life and patient-reported outcomes (PROs).

Objectives: To assess how the change in Pulmonary Function Test (PFTs) parameters correlates with the Patient Reported Outcomes (PROs) in an observational cohort of patients with Systemic Sclerosis (SSc).

Methods: We conducted a retrospective study of 330 clinic episodes from 121 unselected patients diagnosed with systemic sclerosis according to EULAR/ACR 2013 criteria, in annual follow-up (for a total of 165 patients/year) with PFTs, Health Assessment Questionnaire Disability Index (HAQ-DI), Scleroderma Health Assessment Questionnaire (sHAQ), Modified Borg Dyspnea Scale (Borg) and Cochin Hand Function Score (CHFS). We assessed the correlation between the HAQ and the Visual Analogical Scale 1-7 at baseline (VAS1 pain, VAS2 disease severity, VAS3 arthritis activity, VAS4 intestinal problems, VAS5 dyspnea, VAS6 Raynaud's phenomenon, VAS7 digital ulcers). We evaluated the correlation of PFTs with PROs at every time period and the correlation between the change of PFTs parameters (δ FVC, δ DLCO) with the change of the PROs over a year of follow-up. Following analysis of distribution, Spearman or Pearson Test were used to determine correlation coefficients, as appropriate (Prism 7).

Results: The median disease duration was 5 years (IQR 3-10). The median of 12 months δ FVC% and δ DLCO% were 0 (IQR -5.81 to 3.28) and -2.439 (IQR -8.76 to 5.98), respectively. The analysis evidenced a strong positive correlation between VAS1-7 and HAQ. We observed also significant correlation between FVC%, DLCO% and HAQ-DI ($r = -0.355$ and -0.266 , respectively; $p < 0.0001$ for both), Borg ($r = -0.403$ and -0.379 , respectively; $p < 0.0001$) and CHFS ($r = -0.355$ and -0.256 , respectively; $p < 0.0001$). Nevertheless, in longitudinal setting there was no significant correlation between δ PROs and changes lung function, as continuous variables, neither there was any significant PROs difference in patients that did or did not lose more than 10% of FVC and DLCO over a year of follow-up.

Conclusion: This analysis of a monocentric non-selected population evidenced that the current commonly used PROs in SSc while showing a good correlation with lung function are poorly sensitive to change or to reflect changes in lung function over 12 months. In this sense, prudent interpretation of the lack of correlation between FVC and patient-reported outcomes in studies of phase 3 is warranted.

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SAT0322

PREVALENCE AND RISK FACTORS FOR LEFT VENTRICULAR DIASTOLIC DYSFUNCTION IN SYSTEMIC SCLEROSIS: RESULTS FROM RESCLE REGISTRY

A. González¹, J. L. Patier¹, M. López-Rodríguez¹, A. Guillén del Castillo², M. Rubio-Rivas³, A. Argibay⁴, B. Mari-Alfonso⁵, A. J. Chamorro⁶, A. B. Madroño-Vuelta⁷, E. L. Callejas-Moraga⁵, C. González-Echávarri⁸, N. Ortego⁹, V. Fonollosa-Pla², C. P. Simeón-Aznar², O. B. O. R. I. Autoimmune Diseases Study Group (Geas)¹⁰ on behalf of RESCLE Investigators, Autoimmune Diseases Study Group (GEAS). ¹Hospital Universitario Ramón y Cajal, Department of Internal Medicine, Madrid, Spain; ²Hospital Universitario Vall d'Hebron, Unit of Autoimmune Diseases, Department of Internal Medicine, Barcelona, Spain; ³Hospital Universitario de Bellvitge-IDIBELL, Unit of Autoimmune Diseases, Department of Internal Medicine, L'Hospitalet de Llobregat, Barcelona, Spain; ⁴Complejo Hospitalario Universitario de Vigo, Unit of Systemic Autoimmune Diseases and Thrombosis, Department of Internal Medicine, Vigo, Pontevedra, Spain; ⁵Parc Taulí, Hospital Universitario, Department of Internal Medicine, Sabadell, Barcelona, Spain; ⁶Hospital Clínico Universitario de Salamanca-IBSAL, Department of Internal Medicine, Salamanca, Spain; ⁷Hospital General San Jorge, Department of Internal Medicine, Huesca, Spain; ⁸Biocruces Bizkaia Health Research Institute, Hospital Universitario Cruces, University of the Basque Country, Autoimmune Diseases Research Unit, Department of Internal Medicine, Bilbao, Spain; ⁹Department of Medicine, Facultad de Medicina, Hospital Universitario San Cecilio, Inst Invest Biosanitaria Ibs Granada, Department of Internal Medicine, Unit of Systemic Autoimmune Diseases, Granada, Spain; ¹⁰Sociedad Española de Medicina Interna (SEMI), Madrid, Spain

Background: Left ventricular diastolic dysfunction (LVDD) is a very common finding in heart involvement in Systemic Sclerosis (SSc).

Objectives: To determine the prevalence, risk factors and mortality associated with LVDD in a cohort of patients with SSc.

Methods: A retrospective study was conducted with data from the multicentre Spanish Scleroderma Registry (RESCLE). A case-control study was performed to identify factors associated with LVDD.

Results: Out of 1517 cases of SSc, 319 (21%) developed LVDD. Basal characteristics are shown in Table 1. In multivariate analysis, LVDD was associated to older age at diagnosis of SSc [54 vs 44 years, OR 1.05 (1.04-1.06)], presence of telangiectasia [67 vs 59%, OR 1.42 (1.88-1.08)], and treatment with calcium channel blockers [50 vs. 45%, OR 1.55 (1.16-1.96)], and inversely correlated to treatment with ACE inhibitors [74 vs. 83%, OR 0.59 (0.44-0.8)]. Mortality was increased in patients with LVDD (24 vs 17%, OR 1.4, $p = 0.01$). Kaplan-Meier cumulative survival for the SSc cohort, according to the presence or absence of LVDD showed significant differences in 30 years from the first SSc symptom (59 vs. 70%, $p = 0.04$).

Table 1.

Number of patients	1517	319 (21%)	1198 (79%)	P
Limited SSc	923 (61%)	201 (63%)	722 (60%)	0.438
Diffuse SSc	304 (20%)	51 (16%)	253 (21%)	0.041
Sine Scleroderma SSc	174 (11%)	47 (15%)	127 (11%)	0.048
Age at disease onset (years) med±SD	46.4±16.5	54.1±15.9	44.5±16.1	<0.001
Arterial hypertension	491 (33%)	148 (48%)	343 (30%)	<0.001
Digital ulcers	615 (41%)	108 (34%)	507 (42%)	0.006
Telangiectasia	918 (61%)	212 (67%)	706 (59%)	0.011
Interstitial lung disease	645 (43%)	153 (48%)	492 (41%)	0.021
Pulmonary hypertension	139 (9.2%)	34 (11%)	105 (8.8%)	0.325
Capillaroscopy Slow pattern	673 (51%)	130 (46%)	543 (53%)	0.038
Centromere Antibodies	679 (49%)	154 (55%)	525 (48%)	0.032

Conclusion: In our cohort, LVDD is relative common in SSc patients, and it is associated with older age at diagnosis, treatment with calcium channel blockers and telangiectasia. ACE inhibitors could play a protective role against the development of LVDD. Mortality is higher in SSc patients with LVDD, and is more significative over time.

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SAT0323

MYOSITIS-SPECIFIC AND MYOSITIS-ASSOCIATED AUTOANTIBODIES IN A LARGE INDIAN COHORT OF INFLAMMATORY MYOSITIS REVEAL NOVEL CLINICO-PHENOTYPIC PATTERNS

L. Gupta¹, P. Gaur², V. Agarwal¹, R. Aggarwal³, R. Misra⁴ on behalf of -
¹Sanjay Gandhi Postgraduate Institute of Medical Sciences, Lucknow, India; ²Niruj Rheumatology Clinic, Ahmedabad, India; ³UPMC, Pittsburgh, United States of America; ⁴Kalinga Institute of Medical Sciences (KIMS), Bhubaneswar, India

Background: Idiopathic Inflammatory Myositis (IIM) are heterogenous, with distinct autoantibodies reflecting upon possible clinical evolution and outcomes. Ethnicity has major influence on both antibody prevalence patterns as well as phenotypic behaviours linked to them.

Objectives: Thus we sought prevalence and co-existence of myositis specific autoantibodies (MSAs) and myositis associated autoantibodies (MAAs) and associated clinical characteristics in a large cohort of patients with IIM.

Methods: Adult patients with a physician diagnosis of IIM as per ACR/EULAR classification criteria were investigated for the presence of MSAs/MAAs by Line immunoassay (G4, Euro-Immune, Lubeck, Germany). Anti-Nuclear Antibody (ANA) was tested by Immunofluorescence assay (IFA), and patterns in various antibody subsets explored. Prevalence and associations of different autoantibodies were assessed in disease subsets and clinical phenotypes.

Results: MSA and MAAs were tested in 250 IIM patients (F:M 3.8:1) of median age 37 (25-47) and disease duration 6 (3-17) years. Dermatomyositis (DM) was seen in most patients 83 (33.2%) followed by overlap myositis (OM), juvenile DM, Anti-synthetase syndrome (ASS), polymyositis (PM), and cancer associated myositis (CAM). MSAs/MAAs were found in 148 (59.2%) of patients, of which 95 (64.2%) had an MSA and 53 (35.8%) had MAAs (Fig. 1A). 93 (62.8%) of autoantibody positive patients were positive for a single antibody, and only 2 (0.8%) of total had more than one MSA (Table 1).

Table 1. Multiple antibodies positive upon testing for MSA and MAA by the LIA

AutoAntigen	Auto-Ab (n)	None	Jo-1	PL-7	PL-12	MDA5	SAE	MI2	NXP2	TIF1y	SRP	Ku	Pm/Scl	Ro52
Jo-1**	21	3			1								1	17
PL-7**	4	1											1	1
PL-12**	7	0	1										1	5
MDA5*	14	8								1*				1
SAE*	5	3						1*						1
MI2	19	17					1							1
NXP2	9	8										1		1
TIF1y	12	9				1								2
SRP**	5	0											1	4
Pm/Scl	12	11		1										1
Ro52	72	33	17	1	5	5	1	1		2	4	3		3
Ku	10	6												3

Note: ** PL-7 co-exists with Ku + Pm/Scl, **PL-12 co-exists with Pm/Scl + Ro52, **SRP co-exists with Pm/Scl + Ro52

The most frequently detected MSA was anti-Jo-1 (8%), with a further 9 specificities each found in 0.5–7.0% of patients. Amongst the autoantibody positive patients, 21% (n=53) had isolated MAA positivity, anti-Ro52 (33, 62.3%) being the most common, followed by anti-Pm/Scl (11, 20.8%) and anti-Ku (9, 17.0%) (Fig. 1B).

Figure 1. A. MSA and MAA in Indian cohort of myositis B. ANA patterns in myositis C. MSA in ANA negative IIM

On ANA, 76.0% (172 of 226) were positive, with speckled being the most common pattern (37%, Fig. 1C). Of those ANA negative (n=54), 61% had either MSA or MAA (Fig. 1D). 18 (54.6%) had autoantibodies associated with cytoplasmic patterns suggesting that cytoplasmic ANA may be underreported. Clinical presentation akin to DM was seen with all MSA except anti-SRP. PM group was heterogenous, and included ASS, OM and necrotizing phenotype (Fig. 2A). On occasion, anti-SRP, anti-Mi-2 and anti-MDA5 presented with clinical phenotype of ASS. (Fig. 2A,C). Patients with ARS or anti-SAE were often clinically amyopathic (Figure 2 B,C)

Figure 2. A. Phenotypic associated with various antibody subsets B,C and D. MSA/MAA in muscle weakness, rash and ILD phenotype. E. Unique feature of eye-lid edema in some patients with MDA-5 positive myositis

ARS were associated with mechanic's hand ($p < 0.0001$, OR 7.6), ILD ($p < 0.0001$, OR 4.4), and arthritis ($p = .002$, OR 2.6) though there was no difference between Jo-1 and non-Jo-1 ASS. Anti-MDA-5 associated with fever ($p = 0.003$, OR 12) and weight loss ($p = 0.008$, OR 10.2) and unique phenotype of eye-lid edema in some adults (Figure 2E) and arthritis in children ($p = 0.01$, OR 11.5). Anti-TIF-1y associated with alopecia ($p = 0.007$, OR 5.9) and malignancy ($p = < 0.0001$, OR 34) in adults but not children.

Conclusion: Myositis autoantibodies are seen in two-thirds IIM and identify distinct clinical subsets as well as unique phenotypes. MSA/MAA are positive in two-thirds of those negative on ANA, adding diagnostic value. MSAs are nearly always mutually exclusive and thus useful as biomarkers for diagnosis.

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SAT0324

SEXUAL HEALTH IN WOMEN AND MEN WITH SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

B. Heřmáková¹, M. Špiritović¹, H. Smucrova², S. Oreska^{2,3}, H. Štorkánová^{2,3}, K. Bubova^{2,3}, K. Pavelka^{2,3}, J. Vencovsky^{2,3}, L. Šenolt^{2,3}, R. Bečvář^{2,3}, M. Tomčík^{2,3}. ¹Faculty of Physical Education and Sport, Charles University, Department of Physiotherapy, Prague, Czech Republic; ²Institute of Rheumatology, Prague, Czech Republic; ³1st Faculty of Medicine, Charles University, Department of Rheumatology, Prague, Czech Republic

Background: Systemic sclerosis (SSc) is a chronic multisystem disease that may affect all aspects of life including sexual function.

Objectives: To assess sexual function, pelvic floor function in 80 women and 14 men with SSc compared to age-/sex-matched healthy controls (HC) and to analyze the potential impact of clinical features on sexual health.

Methods: In total 80 women (mean age: 49, disease duration: 6.1 years, lcSSc/dcSSc: 53/27, mRSS: 9.3, ESSG activity index: 2.1) and 14 men with SSc (mean age: 51.6, disease duration: 3.1 years, lcSSc/dcSSc: 6/8, mRSS: 12.9, ESSG activity index: 2.2) who fulfilled the ACR/EULAR 2013 criteria, filled in 12 well-established and validated questionnaires assessing sexual function, pelvic floor function, quality of life, fatigue, physical activity and depression. Full names of questionnaires are listed in Table 1. These questionnaires were also completed by 80 healthy women and 14 healthy men with identical age. Data are presented as mean±SEM.