Disclosure of Interest: H. Maki: None declared, M. Hatano Grant/research support from: Ministry of Health, Labour and Welfare, Bayer Japan, Nippon Shinyaku Co,.Ltd, GlaxoSmithKline K.K., Speakers bureau: Actelion pharmaceuticals Japan Ltd, Bayer Japan, S. Minatsuki: None declared, T. Inaba: None declared, I. Komuro Grant/research support from: Actelion pharmaceuticals Japan Ltd, Y. Asano Grant/research support from: Ministry of Health, Labour and Welfare, S. Sato Grant/research support from: Ministry of Health, Labour and Welfare
DOI: 10.1136/annrheumdis-2017-eular.4271

AB0636 A COMPARISON STUDY OF PREVALENCE OF TRADITIONAL CARDIOVASCULAR RISK FACTORS AND FRAMINGHAM RISK SCORE IN SYSTEMIC SCLEROSIS PATIENTS AND MATCHED CONTROLS

J. Sousa-Neves, M. Cerqueira, D. Santos-Faria, J. Leite Silva, C. Afonso, F. Teixeira. *Rheumatology, Hospital Conde de Bertiandos, ULSAM, Ponte de Lima, Portugal*

Background: In Systemic Sclerosis (SSc), data on prevalence of traditional cardiovascular (CV) disease risk factors is scarce and conflicting (1). Therefore, SSc patients CV risk attributed to traditional CV risk factors remains an issue of debate.
Objectives: To evaluate if patients with SSc have a higher prevalence of traditional CV disease risk factors and a higher risk of long-term CV events based on the risk prediction tool of the Framingham risk score (FRS) in comparison with age, race and sex matched control subjects.
Methods: The study comprised patients diagnosed with SSc, fulfilling both the 1980 ACR and the 2013 ACR/EULAR criteria for the disease, and followed-up at our Rheumatology Department and a group of age, race and sex-matched controls. Inclusion criteria were age 30 to 74 and no history of CV events in order to calculate FRS. In total, 46 out of 62 patients were eligible for the study. Traditional CV disease risk factors (diabetes, arterial hypertension and smoking) were compared among the 46 patients with SSc and 51 matched controls. Systolic blood pressure (SBP) values and total and high-density lipoprotein (HDL) cholesterol levels were also collected. The 10-year risk for CV events according to FRS was calculated and means of patients and controls were compared. Subjects' distribution into 3 categories of risk – low (<10% risk), medium (10–20% risk) and high (>20% risk) was also compared. Parametric and nonparametric tests were used for comparison between groups. P value <0.05 was defined as statistically significant.
Results: Mean risk for CV events in 10-years assessed by FRS was 10.00%±8.61 for SSc patients and 7.76%±8.30 for matched controls. Differences were not statistically significant (p=0.196). Additionally, prevalence of diabetes, arterial hypertension and smoking did not differ significantly between the two groups (p=0.890, p=0.443, p=0.651, respectively). Total and HDL cholesterol levels were also similar between groups (p=0.963 and p=0.506, respectively). Only SBP values (mmHg) of SSc patients were significantly higher (128.50 mmHg [113.5 to 139.3]) (median [interquartile range]) compared with controls (120.00 [110 to 130]), p=0.031. Subjects' distribution into the 3 groups of risk defined was similar for both groups (p=0.205).
Conclusions: In our study, prevalence of traditional CV disease risk factors and 10-year risk for CV events based on FRS assessment tool did not differ significantly between SSc patients and age, sex and race matched controls.
References:
[1] Psarras A, Soulaidopoulos S, Garyfallos A, Kitas G and Dimitroulas T. A critical view on cardiovascular risk in systemic sclerosis. Rheumatol Int. 2017 Jan;37(1):85–95.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.4528

AB0637 SUBCLINICAL ATHEROMATOSIS AND VITAMIN D DEFICIENCY IN PATIENTS WITH SCLERODERMA

J.J. Gonzalez Martin[1], O. Carrion[2], A. Abdelkader[2], E. Calvo[1], F. Aramburu[1], F. Sainz[2], P. García de la Peña[1]. *[1]Rheumatology; [2]Angiology and Vascular Surgery, Hospital Universitario HM Sanchinarro, Madrid, Spain*

Objectives:
To study whether patients with systemic sclerosis (SSc) have an increased cardiovascular risk (CVR), measured on the basis of analytical, angiodinamic and/or vascular lesions on carotid ultrasound.
The carotid IMT is a marker of cardiovascular morbidity and mortality, allowing measurement and monitoring of atherosclerosis in asymptomatic individuals, being surrogate markers of future coronary disease, stroke and general death in the general population and in inflammatory rheumatologic diseases.
Methods: Epidemiological and analytical data were collected, including the determination of the RCV SCORE index.
Vascular ultrasound protocol included assessment of carotid intima-media thickness (IMT), presence of atheromatous plaques, and exploration of peripheral arteriopathy using the ankle arm index (ABI).
Results: Seventy adult patients with ES diagnosis (ACR-EULAT 2013 criteria) were included.
94% of the women had a mean age of 50.2±12.5 years, and an average evolution time of 3.0±4.4 years.
The distribution by subgroups was: limited SSc (48%), diffuse SSc (34%), pre-SSc (4'2%), sine SSc (2.8%), MCTD (5'7%) and overlap syndrome 4'2%). The mean SSRm was 9.3±7.0 (range 0–42).
The ANA were positive in 91.4%, ACA (51.4%), ATA (10%), RNA polymerase (4'2%).
4% were DM, 7% were obese, 11% were active smokers, 13% were HTN, and 28% were ex-smokers.
28% had hypercholesterolemia with a mean total cholesterol of 192.5 (SD ± 31.9) and LDL of 102.4 (SD ± 29.4 mg/dL).
57% received vasodilators, most of them ARA-II. 10% bosentan, 4.2% sildenafil, and a 2.8% combination therapy.
The percentage of immunosuppressive drugs was corticoid (50%), MTX (34%), mycophenolate (3%), AZA (11%), HCQ (14%), CP %).
The IMT presented pathological values (>0.9 mm) in 39% of the sample, 23% had atheroma plaques (being bilateral in 40%). Subclinical atheromatosis affected 41.4% (patients without cardiovascular events, pathological IMT and/or atheroma plaques). The ABI had pathological values (<0.9) in 17% of the patients.
In the bivariate analysis, the pathological GIM was related to the presence of ACA antibodies (OR =3.80, 95% CI: 1.15–12.52, p=0.028) and with the SCORE index of CVR (OR =2.93, 95% CI: 1.12–7, 64, p=0.028); And the presence of atherosclerotic plaques was associated with increased SSRm score (OR 1.09, 95% CI 1.00–1.19, p=0.046), and the highest CVR SCORE index (OR 3.90, 95% CI: 1.31–11.56, p=0.014.
In the multivariate analysis, the serum vitamin D concentration showed a protective effect on IMT (OR =0.94, 95% CI 0.89–0.99, p value =0.025); And the main determinant of atheromatous plaques is the SCORE index, since the increase of one unit in SCORE index multiplies by 4 the probability of presenting plaques (OR =4.06, 95% CI: 1.31–12.60; P=0.015), once the effect of SSRm was controlled.
Conclusions:
- 40% of the patients had pathological IMT values, showing association with the presence of positive AAC and the SCORE risk index.
- The serum concentration of 25-OH-vitamin D showed a protective effect on IMT. Sixty percent of the sample had vitamin D deficiency.
- The presence of atheromatous plaques (23% of patients) was associated with higher SSRm indexes and SCORE cardiovascular risk.

Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6995

AB0638 CARDIAC TRANSPLANT IN SYSTEMIC SCLEROSIS-ASSOCIATED CARDIOMYOPATHY: MONOCENTRIC EXPERIENCE OF 3 CASES

J. Lenaerts[1], J. Lenaerts[2], R. Westhovens[2], W. Droogne[3], A. Ciarka[3], J. Van Cleemput[3], E. De Langhe[2]. *[1]Internal Medicine; [2]Rheumatology; [3]Cardiology, UZ Leuven, Leuven, Belgium*

Background: Cardiac involvement in systemic sclerosis (SSc) is a frequent complication, but end-stage cardiac failure remains uncommon and represents a poor prognosis. Heart-lung and lung transplant is an established treatment option for SSc-related pulmonary disease. Due to the limited published data, no recommendations exist for cardiac transplant in the context of SSc.
Objectives: We present our monocentric experience of 3 patients with SSc who underwent cardiac transplant for SSc-related end-stage heart disease (multiple hospitalisations due to failure of medical therapy and life-threatening complications).
Results: *Case 1* is a 59-year-old male with limited cutaneous SSc. Antinuclear antibody (ANA) was negative. He had vascular (digital ulcers) and cardiac (heart failure (left ventricular ejection fraction (LVEF) 20%, NYHA class IV)) involvement, without major gastrointestinal or pulmonary involvement (no interstitial lung disease (ILD) or pulmonary arterial hypertension (PAH: assessed by right heart catheterization (RHC))). He underwent a cardiac transplant at the age of 51, after a disease duration of 6 years. Post-transplantation immunosuppressant therapy consists of tacrolimus and mycophenolic acid, initially associated with methylprednisolon, which is the standard immunosuppression protocol at our institution.
Case 2 is a 55-year-old male with limited cutaneous SSc. ANA was positive,

	Case 1	Case 2	Case 3
Sex, age	Male, 59 years	Male, 55 years	Male, 50 years
Type of SSc	Limited	Limited	Diffuse
Disease duration at Tx	6 years	7 years	4 years
ANA	Negative	1/320, speckled, no SSc-specific antibody	Negative
Pre-Tx			
NYHA class	IV	III	III
LVEF	20%	40%	40%
RHC: mPAP in mmHg	26	20	31
RHC: PCWP in mmHg	20	14	28
Post-Tx			
NYHA class	I	I	I
LVEF	60%	55%	60%
RHC: mPAP in mmHg	19	13	21
RHC: PCWP in mmHg	12	8	12

Tx: Transplant; mPAP: Mean pulmonary artery pressure; PCWP: Pulmonary capillary wedge pressure.

1/320, speckled pattern, but no SSc-related autoantibody has been identified. He had gastrointestinal (upper gastrointestinal tract dysmotility), muscular (myositis) and cardiac (heart failure with secondary cardiac cirrhosis (LVEF 40%, NYHA class III)) involvement, without major pulmonary involvement (no ILD or PAH). He underwent a cardiac transplant at the age of 54, after a disease duration of 7 years. Standard immunosuppressants were initiated.

Case 3 is a 50-year-old male with diffuse cutaneous SSc. ANA was negative. He had vascular (digital ulcers), gastrointestinal (upper gastrointestinal tract dysmotility) and cardiac (heart failure with secondary cardiac cirrhosis (LVEF 40%, NYHA class III)) involvement, without major pulmonary involvement (no ILD or PAH). He underwent a cardiac transplant at the age of 49, after a disease duration of 4 years. Standard immunosuppressants were initiated.

At present, 1,5 years (case 2 and 3) and 8 years (case 1) after transplant, the donor hearts are still functioning well. No other SSc-related organ manifestations have occurred.

Conclusions: We present 3 patients with SSc who successfully underwent cardiac transplant for SSc-related end-stage heart disease. None had other major SSc-related organ involvement. This supports the limited published data that cardiac transplant is feasible and can be considered in end-stage SSc-related cardiomyopathy.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.4251

AB0639 SUPERIOR TREATMENT RESPONSE OF INTERSTITIAL LUNG DISEASE IN INFLAMMATORY MYOPATHIES COMPARED TO OTHER CONNECTIVE TISSUE DISEASES – A PROSPECTIVE COHORT STUDY

J.J. Maya[1], A. Borja[2], A. Abril[1], I. Mira-Avendaño[2]. [1]*Rheumatology;* [2]*Pulmonary, Mayo Clinic Florida, Jacksonville, United States*

Background: Interstitial Lung Disease (ILD) associated to Connective Tissue Disease (CTD) represent a challenge for clinicians and researchers because of their significant morbidity and mortality. Although the different types of ILD associated to CTD are often studied and managed as one because of their autoimmune background, there are considerable differences in their etiopathogenesis and therefore it can be assumed that there are differences in their response to treatment (1). Even though previous studies have analyzed the impact of immunosupipression in ILD secondary to sclerodema, additional studies are needed in order to determine the response to treatment of the different forms of ILD associated to CTD (2–3).

Objectives: To characterize and analyze the response to treatment of different types of ILD associated to CTD. The primary endpoint is the Functional Vital Capacity (FVC) change at 6 months and 1 year, and secondary endpoints arere the change in Diffusion Capacity of the Lung for Carbon Monoxide and in a 6 Minute Walk Test.

Methods: A prospective cohort study is being carried out where all patients who present to the Mayo Clinic Florida pulmonary clinic, age 18 to 80, with established CTD and diagnosed with ILD, and all patients with ILD who meet the criteria for immunologic mediated process, are being followed for a year in order to evaluate the clinical and functional outcomes to treatment. Patients with moderate to severe Pulmonary Hypertension, and active smokers with bronchiolitis pattern are being excluded. Exploratory analysis were performed on the first group of patients enrolled in the study, continuous variables were described with central tendency measures and the mean absolute difference in adjusted 12-month FVC was analyzed between the different types of CTDs using student's t test.

Results: Thirteen patients with ILD were enrolled in the study's initial phase. Five of the patients had been diagnosed with an Inflammatory Myopathy (IM), 2 with Rheumatoid Arthritis, 1 with an Undifferentiated Connective Tissue Disease, 1 with Churg-Strauss Syndrome, and one with Systemic Sclerosis. One patient was treated with Rituximab only; 2 with Rituximab and a steroid; 3 with Mycophenolate Mofetil (MMF) only; 2 with steroids, MMF, and Rituximab; 1 with a TNF inhibitor and MMF; 1 with MMF and steroids; 1 with Azathioprine and steroids; and 1 received only steroids. IMs were compared to the rest. At 1 year follow-up FVC mean absolute difference for the IMs demonstrated and improvement of 0.43 while the other CTDs had worsen by a mean of 0.04 (p=0.01). There were no statistical differences at 6 months for all outcomes or at 1 year for DLCO and 6MWT.

Conclusions: To our knowledge, this is the first time that a cohort of ILD associated to CTD patients is followed and analyzed after 1 year of treatment. Our results suggest that there is a difference in the response to treatment of ILD depending on the underlying CTD. Specific types of ILD associated to CTD likely benefit from early and aggressive treatment, but longer follow ups and larger studies are needed in order to be able to validate and further understand these pathologies.

References:

[1] Chest 2013; 143 (3): 814–824.
[2] Lancet Respir Med. 2016 Sep;4(9):708–19.
[3] Semin Arthritis Rheum. 2016 Oct 13.[Epub ahead of print].

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.5653

AB0640 DOES MIXED CONNECTIVE TISSUE DISEASE WITHOUT ANTI-U1RNP EXIST?

J. Martínez-Barrio[1,2], E. Estrada[3,4], J.G. Ovalles-Bonilla[1], L. Valor[1], D. Hernández-Flórez[1], T. del Río[1], I. Janta[1], J.C. Nieto Gonzalez[1], B. Serrano[1], R. González Benítez[1], C. Sáenz Tenorio[1], L. García-Montoya[1], M. Correyero[1], A. Silva[1], A. López-Cerón[1], C. Gonzalez[1], I. Monteagudo[1], F.J. López-Longo[1]. [1]*Department of Rheumatology, Hospital General Universitario Gregorio Marañón;* [2]*Universidad Complutense de Madrid;* [3]*Facultad de Salud, Universidad Camilo José Cela;* [4]*Facultad de Psicología, Universidad Autónoma de Madrid, Madrid, Spain*

Background: Mixed Connective Tissue Disease (MCTD) is a systemic autoimmune rheumatic disease (SARD) characterized by clinical manifestations of systemic lupus erythematosus (SLE), systemic sclerosis (SSc) and polymyositis (PM) and the presence of anti-U1-RNP antibodies.

Objectives: To determine whether there are patients with symptoms of MCTD in the absence of anti-U1-RNP antibodies.

Methods: This was a monocentric, prospective, observational study of patients with SARD. All patients diagnosed of MCTD according to Kasukawa and/or Alarcón-Segovia's criteria, SLE, SSc, PM, overlap syndromes (simultaneous or sequential criteria of 2 or more SARD), Sjögren's syndrome, Antiphospholipid syndrome, systemic vasculitis and undifferentiated or incomplete SARD (at least one clinical criterion of the classification criteria and a related antibody of any of the SARD) were included in the "Autoimmune Systemic Rheumatic Diseases Registry" of the Hospital General Universitario Gregorio Marañon Rheumatology Department from 1986 to 2012. The registry includes 2406 patients diagnosed with SARD. Patients with rheumatoid arthritis were excluded. Patients with clinical MCTD criteria were divided into seropositive (MCTD, with anti-U1RNP) and seronegative (possible MCTD, without anti-U1RNP). The registry counts with the local Institutional Ethics Board approval.

Results: A total of 692 patients were recruited, 608 women (87.9%). Seventy (70, 10.1%) patients were classified as seropositive and 75 (10.8%) as seronegative by Kasukawa's criteria. Sixty-two (62, 8.9%) patients were classified as seropositive and 54 (7.8%) as seronegative according to Alarcón-Segovia's criteria. There were no significant differences in age at disease onset, age at diagnosis or disease duration ($p>0.05$) between seropositive and seronegative patients. Seropositive patients with Kasukawa's criteria presented more frequently: lymphadenopathy, malar rash, leukopenia, Raynaud's phenomenon, muscle weakness and increase of muscle enzymes (Table 1). By Alarcón-Segovia's criteria, patients who developed myositis were more frequent in the seropositive group (p=0.007, OR 3.25, 95% CI, 1.44–7.32).

Kasukawa criteria	Seropositive MCTD		Seronegative MCTD		*P*	OR	95% CI OR	
	n	%	*N*	%			Inf	Sup
Lymphadenopathy	28	40%	17	23%	0.038	2.275	1.105	4.681
Malar rash	25	36%	8	11%	0.001	4.653	1.928	11.231
Leucopenia	41	59%	17	23%	<0.001	4.824	2.348	9.909
Muscle weakness	32	50%	24	32%	0.042	2.125	1.083	4.171
Increase of muscle enzymes	45	64%	35	47%	0.049	2.057	1.056	4.008

Conclusions: Some patients with SARD manifestations fulfill MCTD clinical criteria, both Kasukawa's and Alarcón-Segovia's, in the absence of anti-U1-RNP antibodies from the onset of the disease and throughout its evolution (seronegative MCTD). The frequency of seronegative MCTD was similar to the frequency of seropositive MCTD. Patients with seropositive MCTD presented more frequently manifestations of SLE (lymphadenopathy, malar rash and leukopenia) when using Kasukawa's criteria and of PM when using both criteria.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.5192

AB0641 ARTERIAL STIFFNESS AND CLINICAL ASSOCIATION IN PATIENTS WITH SYSTEMIC SCLEROSIS

K. De La Cruz Rodríguez[1], G. Martínez Bonilla[2], V. González Díaz[2], K. García Osuna[2], J. Reyes Rueda[2], C. Gómez López[2], J. Polanco Cruz[2], A. Bernard Medina[2], E. Cardona[3], A. Macías Chumacera[4], C. Ramos Becerra[4], S. Gutiérrez Ureña[2], S. Cerpa Cruz[2]. [1]*Rheumatology Service;* [2]*Hospital Civil Fray Antonio Alcalde;* [3]*Mechanical Vascular Service;* [4]*CUCS, Guadalajara, Jalisco, Mexico*

Background: Systemic sclerosis is an autoimmune disease characterized by microvascular damage and fibrosis. There are several studies that shown macrovascular damage with arterial stiffness (AS) and the risk of cardiovascular complications. Carotid-femoral pulse wave velocity (CF-PWV) and augmentation index (AIx) are two competent methods to determine AS and predictors of cardiovascular disease. Association between AS and microvascular damage is unknown in systemic sclerosis patients.

Objectives: To determine the frequency of arterial stiffness in patients with systemic sclerosis and its association with clinical manifestations.

Methods: We performed a cross-sectional study; patients with diagnosis of systemic sclerosis according to ACR/EULAR 2013 criteria were included and the control group was selected from a database of mechanical vascular service. AS