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

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

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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].

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

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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.

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AB0641 ARTERIAL STIFFNESS AND CLINICAL ASSOCIATION IN PATIENTS WITH SYSTEMIC SCLEROSIS

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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