a significant difference was found between the other groups ($p < 0.05$). IHC with YKL-40 revealed staining of the vessels in the superficial dermis in SSc and normal skin remained unstained. The ROC curve for PAH showed an AUC=0.959, and the most valid cut-off levels of YKL-40 age percentile was 85.2 (sensitivity 86.7% and specificity 92.8%).

Conclusions: Our report is the first report of IHC with YKL-40 in SSc. IHC suggest that YKL-40 may be further elevated by angiogenesis caused by microcirculatory damage in SSc. Our results suggest that YKL-40 may be very useful biomarker for diagnosing SSc and complications associated with microvascular lesions.

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AB0675 ANTI-MDA5 POSITIVE DERMATOMYOSITIS: AN EMERGING ENTITY

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Background: Hypo or amypathic dermatomyositis (DM) represent about 20% of the classic DM. Melanoma differentiation-associated gene 5 antibody or anti-MDA5 antibody, formerly known as anti-CADM140 antibody, has been reported in the last decade. This antibody is detected in more than 50% of patients with hypo or amypathic DM and it has specific features such as the presence of ulcerative vascular lesions and rapid progression of the interstitial lung disease.

Objectives: To report anti-MDA5 positive DM cases diagnosed at a university county hospital, between 2012 and 2016.

Methods: Design: Retrospective descriptive study. Location: University county hospital. Reference area: ~350.000 inhabitants. Clinical manifestations, analytical results, diagnostic approach, treatment administered and follow-up have been exposed.

Results: We report four anti-MDA5 positive DM with a mean age at diagnosis of 54 years (35–77). Caucasian origin in 2 men and Maghreb origin in 2 women. They had no relevant medical history neither neoproliferative process was detected.

ANA were negative and CK values were normal in all the cases. Nailfold capillaroscopy was characteristic of DM pattern. Case B did not present cutaneous manifestations and he was diagnosed as anti-MDA5 DM after his death.

Discussion: B and C cases showed abnormal heart rhythm that could be related to myocardial involvement, as suggested in a recent publication of anti-MDA5 DM (1). In case C, sinus tachycardia was resolved with the clinical response to treatment.

In the literature reviewed, anti-MDA5 positive DM has been reported as a dermatomyositis syndrome, but probably the clinical profile of these patients remains to be defined. In our cases, one of them did not present skin lesions and another case did not develop pulmonary involvement 18 months after the diagnosis.

Conclusions: Keep in mind anti-MDA5 antibody in patients with cutaneous manifestations of DM with vasculopathy lesions, minimal or absent muscular disease, heart rhythm abnormalities not explained by other causes and/or rapidly progressive pulmonary involvement.

Anti-MDA5 positive DM may have a poor response to treatment and fatal outcome. It remains to be seen if early diagnosis could improve the life expectancy of these patients.

Abstract AB0675 – Table 1

	A	B	C	D
Sex	Female	Male	Female	Male
Age at disease onset	43	77	35	62
Fever	Yes	No	No	No
Heliotrope rash	Yes	No	Yes	No
Gotttron papules	Yes	No	No	No
Skin ulceration	No	No	No	Yes
Joint disease (J)	No	No	RA like	Palindromic like
Raynaud's disease	No	No	No	No
Muscular disease (M)	No	No	Yes	No
Pulmonary disease (P)	Bronchiolitis	Interstitial lung disease	No	Interstitial lung disease
Cardiac disease (Card)	No	Arrhythmias	Sinus tachycardia	No
Aldolase (N<7)	Normal	NA	11	14,6
ESR/CRP mg/dL	30/4,5	41/13,7	102/8,1	50/1,8
Ferritin (30–400 ng/ml)	1253	NA	225	2437
Electromyography	NA	NA	Normal	NA
Muscle biopsy	NA	NA	Miopathy	NA
Skin biopsy	DM	NA	Nonspecific	DM
Treatment	CC + CPM + TACR + IVIG	CC	CC + MTX + HCQ	CC + CPM + TACR
Outcome (months)	Death	Death	Clinical response (18 m)	Alive (16 m)

RA = Rheumatoid Arthritis; NA = Not Available; ANA = Antinuclear Antibodies; DM = Dermatomyositis; CC = Corticosteroids; CPM = Cyclophosphamide; TACR = Tacrolimus; IVIG = Intravenous Immunoglobulins; MTX = Methotrexate; HCQ = Hydroxychloroquine.

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AB0676 RELATIONSHIP BETWEEN BIOLOGICAL BIOMARKERS AND CHANGES IN RIGHT VENTRICLE THAT PRECEDE PULMONARY HYPERTENSION IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: A higher prevalence of Pulmonary Hypertension (HP) has been described in patients with Systemic Sclerosis (ES) diagnosis, so this fact significantly overshadows the prognosis, since it leads to the failure of the right ventricle (RV). In this context, it could be very useful to identify ES patients who are at greater risk of developing PH in the future to make an early diagnosis of this complication, and prescribing an early therapy to improve their prognosis.

Objectives: The purpose of this study is to investigate if certain fibrosis, inflammation and vasculopathy biomarkers could be associated to early functional changes in RV, assessed by echocardiography and cardiac nuclear magnetic resonance (MRI) in ES patients.

Methods: A cross-sectional study was performed on a cohort of ES patients diagnosed according to the LeRoy modified criteria, who were being following-up in the Systemic Autoimmune Diseases Unit of the Internal Medicine Service of La Paz University Hospital. Serum levels of inflammation (IL-13, IL-6, TNF alpha), vasculopathy (VEGF), fibrosis biological markers (endoglin, PDRFR, TGFB1, TGFB2, TGFB3) and cardiac biomarkers (NT-proBNP, High sensitivity T troponin) were determined as well as hemodynamic and cardiac morphological variables, were measured by transthoracic echocardiography (systolic and diastolic eccentricity index, E/E' ratio, E/A, pulmonary artery systolic pressure, RV thickness) and cardiac MRI (Mean pulmonary artery velocity). We investigated how many patients had these markers elevated and how many of them had hemodynamic and morphological cardiac altered measurements.

Results: Twenty patients with a diagnosis of ES (18 females/2 males) aged between 41 and 77 years, with a mean of 56 years, were included. A control group of 9 patients with similar demographic characteristics was included. ES subjects had higher levels of PDGFR (fibrosis marker) compared to the control group. In addition, in the group of ES patients, two statistically significant associations were observed: troponin T levels (hs-cTnT), endoglin and TGFB2 had higher levels in patients with a systolic eccentricity index >1; a negative correlation was evidenced between levels of NT-proBNP and the mean pulmonary artery velocity measurement (VELAP).

Conclusions: Hs-cTnT, Endoglin and TGF beta are biomarkers that could appear

elevated in patients with systolic eccentricity index >1 . NT-proBNP is elevated in patients with decreased VELAP. This study demonstrates the association between certain cardiac biomarkers, fibrosis and vasculopathy peptides and early finding suggesting cardiac remodeling that occurs in some ES patients, which may help to predict the development of PH in those patients.

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AB0677 INDIVIDUAL IMMUNOLOGICAL PREDICTORS OF THE RISK OF LUNG DAMAGE IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: A significant role in the formation and development of systemic sclerosis (SSc) belongs to immunogenetic factors and immunopathological mechanisms, so nowadays become important to find immunological predictors of early diagnosis of the disease. An urgent task is timely diagnosing the pulmonary complications as the main causes of high mortality and appointment of adequate therapy and objective assessment of its effectiveness.

Objectives: The main purpose was to develop a personal way of forecasting the risk of damage of the lungs, including its subtype – pneumofibrosis (PF) and pulmonary hypertension (PH) in patients with SSc.

Methods: To address this goal, the following studies performed: clinical, general laboratory, instrumental, immunological, molecular genetics, statistical methods. Adequacy and reliability of the results of mathematical model of the risk of damage the lungs were using criteria Wald and Xi-square. The results show that our model is correct with a probability of error less than 1% ($p=0.001$).

Results: We found that among the analyzed complex interrelated factors in the development of lung damage in patients with SSc significantly influenced: the duration of the disease, miRNA29b, expression levels in the blood concentration of IL17 in serum and blood, neutrophil phagocytic index that we include in the predictive model.

We know that the most significant complications associated with damage to their lungs are pneumofibrosis and pulmonary hypertension, which threatened the rapid development of respiratory failure and a high mortality of these patients. In this regard analyzed immunological and molecular genetic parameters of patients with SSc depending on the development of PF and LH.

We found that among the complex of interrelated factors of the development of PF in patients with SSc significantly influenced the following factors: patient age, miRNA 29b, MCP-1, IFN- γ , TGF- β in serum, oxidative indicator “explosion” of neutrophils, which we have included in the predictive model.

At the same time, the development of PH in patients with SSc significantly affected other clinical and immunological factors: patient age, levels of endothelin-1, IL17 in serum, the number of CD4 + CD25 + Fox P3 + -lymphocytes, CD3 +/CDHLA-DR + -lymphocytes, CD4 + IL17 + -lymphocytes in peripheral blood, which we have included the following predictive model.

Conclusions: The application withdrawn prognostic clinical and immunological and molecular genetic criteria of lungs damage (pulmonary hypertension and pneumofibrosis) will enable medical practitioners to verify early visceral lesions in patients with SSc in time and justified the appointment of basic therapy.

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AB0678 ACCELERATED ACTIVATION OF 25-HYDROXYVITAMIN D TO 1,25-DIHYDROXYVITAMIN D IN SYSTEMIC SCLEROSIS PATIENTS- A RETROSPECTIVE STUDY

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Background: There have been multiple reports of low vitamin D in Systemic

Sclerosis (1, 2). The majority of these reports only measured 25-hydroxyvitamin D (calcifediol) and not 1,25-dihydroxyvitamin D (calcitriol). In our clinical observation, we noted that there may be a population of patients with low calcifediol but normal or high calcitriol.

Objectives: We aim to evaluate the frequency of normal or high calcitriol level in systemic sclerosis patient with low calcifediol levels.

Methods: We performed a retrospective analysis using data collected from a large private practice. We collected data from systemic sclerosis patients who had calcifediol and calcitriol levels ordered between January 2014 and January 2017, patients with sarcoidosis overlap were excluded.

Results: We identified 90 scleroderma patients with documented calcifediol and calcitriol levels and no history of sarcoidosis. Mean calcifediol level was 32.7 ng/mL (SD 20.2) and mean calcitriol level was 60.9 pg/mL (SD 27.6). Total of 42 patients were identified as having calcifediol level of less than 30 (normal >30), only 1 of these patients had calcitriol levels below 10 pg/mL (normal >10) and 6 patients had calcitriol levels above 75 pg/mL. Twenty-three out of 41 patients who had low calcifediol but high calcitriol level also had a CT chest with no diagnostic findings suggestive of sarcoidosis.

Conclusions: Our study suggests that while low calcifediol levels are common in systemic sclerosis patients, not all patients are truly vitamin D deficient and a calcitriol level should be checked before supplementation is initiated. There is some documented evidence of sarcoid and scleroderma overlap at greater than expected frequency which is under recognized and elevated calcitriol level in a systemic sclerosis patient should prompt further evaluation for sarcoidosis (3). We have identified a population of scleroderma patients with low calcifediol, elevated calcitriol level, but no evidence of sarcoidosis. Based on this observation we are planning a prospective study to investigate this further.

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AB0679 THE CLINICAL VALUE OF NAILFOLD CAPILLAROSCOPY IN THE EARLY DIAGNOSIS OF SYSTEMIC SCLEROSIS

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Background: Vascular changes was early pathological changes of SSc, gradually appearing with irreversible fibrosis of the skin and internal organs. Early diagnosis and assessment of the development and efficacy timely, could improve survival in patients with SSc.

Objectives: To assess the value of nailfold capillaroscopy in the early diagnosis of systemic sclerosis (SSc).

Methods: 60 patients with SSc and 55 patients with other connective tissue diseases. Data were extracted on clinical and laboratory parameters. 2013 ACR/EULAR classification criteria and 1980 ACR criteria for SSc were evaluated.

Results: The sensitivity of the 2013 criteria was 91.7% compared to 56.7% for the 1980 criteria ($P<0.001$). The specificity of two criteria was no significant difference. This sensitivity of the 2013 criteria was higher compared to the 1980 criteria among those with lcSSc (95.5% versus 50%). The pattern was consistent among those with disease duration <3 years (90.5% versus 57.1%, $P<0.05$), and disease duration ≥ 3 years (92.3% versus 56.4%, $P<0.05$). The sensitivity and specificity of nailfold capillaroscopy to determine SSc were 86.7% and 43.6%. Patients not fulfilling the two classification criteria were met the very early diagnosis of systemic sclerosis, and often suffering from RP, and had an SSc pattern on nailfold capillaroscopy.

Conclusions: The sensitivity of 2013 ACR/ EULAR classification criteria was higher compared with 1980 ACR classification criteria. The specificity of two classification criteria was no significant difference. This sensitivity of two criteria was higher among those with lcSSc and short disease duration. Scleroderma pattern were significantly associated with the development of systemic sclerosis.

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