Conclusions: In SSc-patients, TA were predominantly located on the face, hands and the upper part of the trunk. They may reflect the vasculopathy of SSc and could represent a clinical biomarker for vascular disease, particularly for PH, one of the most severe vascular complications of the disease.

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THU0418

LONG-TERM EFFICACY AND SAFETY OF MONOTHERAPY VERSUS COMBINATION THERAPY IN SYSTEMIC SCLEROSIS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION (PAH): A RETROSPECTIVE COHORT STUDY FROM THE NATIONWIDE SPANISH SCLERODERMA REGISTRY (RESCLE)

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Background: Monotherapy with endotelin antagonist receptors (ERA) an phosphodiesterase 5 (PDE5) inhibitors is a first choice treatment for PAH in functional class (FC) II-III, with the same grade of evidence and recommendation than combination therapy. Recently, studies have proven superiority of combination therapy against monotherapy in combined morbi-mortality endpoints.

Objectives: To demonstrate superiority of combination therapy against monotherapy in a single mortality endpoint in SSc-associated PAH.

Methods: Retrospective cohort study including patients from the Spanish Scleroderma Registry (RESCLE) diagnosed with SSc-associated-PAH by right heart catheterization (RHC). Patients were divided in 3 groups: monotherapy vs. sequential combination therapy (>12 weeks between first and second treatment)

vs. upfront combination therapy (<12 weeks between treatments). Primary end-point was mortality from any cause.

Results: Seventy-six patients with PAH out of 1817 participants were included. Thirty-four (45%) were receiving monotherapy [with ERA (22 patients, 29%) or PDE5 inhibitors (12 patients, 16%)], 25 patients (33%) sequential combination therapy and 17 patients (22%) upfront combination therapy.

Baseline demographic, clinical and complementary tests were similar among groups. ILD (mainly moderate) was more frequent in both combination groups in 58% vs. 80% vs. 76.4%, without statistical significance. A worse FVC/DLCO in the sequential combination group was reported (2.9 ± 1.1 vs. 1.8 ± 0.4 vs. 2.3 ± 0.8 , global $p=0.085$ but $p=0.043$ comparing monotherapy with sequential combination) and also a worse mPAP in both sequential and upfront combination groups (37.2 ± 8.7 mmHg vs. 40.8 ± 8.8 vs. 46 ± 15.9 , $p=0.026$).

The treatment regimen prescribed ($p=0.017$) and FC at baseline ($p=0.007$) were found predictors of mortality. Sequential combination therapy was found a protective factor [HR=0.11 (95%CI 0.03–0.51), $p=0.004$] and the upfront combination therapy showed a tendency of protection [HR=0.68 (95%CI 0.23–1.97), $p=0.476$]. Survival rates from diagnosis of PAH among groups were: 78% vs. 95.8% vs. 94.1% at 1 year, 40.7% vs. 81.5% vs. 51.8% at 3 years and 31.6% vs. 56.5% vs. 34.5% at 5 years ($p=0.007$).

Side effects were not significantly different among groups.

Conclusions: Combined sequential therapy improves survival in SSc-PAH patients, even with moderate ILD.

Upfront combination therapy may improve survival, but did not reach statistical significance due to study limitations.

Treatment regimen and FC were found as prognostic factors for survival: sequential combination therapy was a protective factor and FC was a risk factor.

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THU0419

ASSOCIATION OF INFLAMMATORY MARKERS C-REACTIVE PROTEIN AND ERYTHROCYTE SEDIMENTATION RATE WITH PULMONARY FUNCTION TESTS AND EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (ESCSG-AI) IN SYSTEMIC SCLEROSIS – ASSOCIATED INTERSTITIAL LUNG DISEASE IN FOLLOW UP STUDY

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Background: inflammatory markers are very important to assess severity and activity of SSc-ILD, but it's role needs further investigation.

Objectives: to assess inflammatory markers of SSc such as hsCRP and ESR and compare with lung function test and ESCSG-AI in the long-term follow up study.

Methods: It was a longitudinal study involving 77 pts with SSc-ILD (mean age was 46.2 ± 13.4 ; 69% have limited subset of the disease; 93% were female). The mean duration of follow up was 58.9 ± 11.4 months. Pts. were investigated with HRCT twice (at first visit and at the end of the study) and according the CT-changes were divided into 3 groups: the 1st group (16 pts) with improvement; 2nd group (39 pts) without any changes and 3rd group (22 pts) with worsening of fibrosis. Other data collected including biological results (high-sensitivity C-reactive protein (hsCRP) and erythrocyte sedimentation rate (ESR)), PFT (forced vital capacity (FVC,% of predicted) and diffusing capacity of the lung for carbon monoxide (DLCO,% of predicted), composite score (ESCSG-AI).

Results: there were no significant differences between groups related to sex, frequency of diffuse form and duration disease. Mean levels of hsCRP and ESR didn't change significantly during the follow up. In all pts the mean levels of hsCRP and ESR correlated directly with each other at first visit and at the end of the study ($R=0.45$ and $R=0.4$ ($p<0.001$) accordingly). We compared the mean levels of hsCRP and ESR with mean dates of FVC, DLCO and ESCSG-AI score in first visit and the end of follow up. Mean levels of hsCRP inversely correlated with mean dates of DLCO at the first visit and at the end of the study ($R=-0.39$ and $R=-0.42$ ($p<0.05$) accordingly); in groups 2 and 3 ($R=-0.34$ and $R=-0.47$ ($p<0.05$) accordingly) at the end of the study; with mean dates of FVC in all pts and group 2 ($R=-0.42$ and $R=-0.47$ ($p<0.05$) accordingly) only at the end of the study; correlated directly with ESCSG-AI score in all pts and groups 2,3 ($R=0.58$ ($p<0.0001$), $R=0.46$ ($p<0.01$) and $R=0.77$ ($p<0.001$) accordingly) at the end of the study. While mean levels of ESR inversely correlated with mean dates of DLCO only in all pts and groups 1,2 ($R=-0.43$, $R=-0.66$ and $R=-0.39$ ($p<0.05$) accordingly) at first visit; correlated directly with ESCSG-AI score in all pts. ($R=0.309$ ($p<0.01$) at the end of the study. Mean levels of hsCRP inversely correlated with DLCO, FVC and directly correlated with ESCSG-AI and these correlations were more evident than with mean levels of ESR.

Conclusions: In our group of pts. the hsCRP has proven to be an accurate reflection of disease severity especially in pts with progression of ILD.

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THU0420

AN EXTENT OF INTERSTITIAL LUNG DISEASE IS A POTENTIAL PREDICTOR OF RESPONSE TO A-B-CELL THERAPY IN THE PATIENTS WITH SSC

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Background: Systemic sclerosis (SSc) is a connective tissue disease associated with chronic polyclonal B-lymphocytic activation and immunological tolerance disturbance. Several research and clinical studies showed that B-cell depletion is potentially efficacious in SSc treatment. However, neither strong evidence of RTX efficacy for treatment of interstitial lung disease (ILD) associated with SSc, no potential predictor of response to a-B-cell therapy.

Objectives: To evaluate rituximab (RTX) therapy efficacy in the patients with systemic sclerosis (SSc) differing in extent of interstitial lung disease (ILD) based on multispiral computed tomography (MSCT) findings.

Methods: 42 patients (average age 48±2 years; male/female 1/6, diffuse/limited disease 1.5/1 (25 and 17), disease duration since the first non-Raynaud syndrome – 6.6±5.9 years) with definitely diagnosed SSc and ILD signs evidenced by MSCT were enrolled into the study. During the observation period 29±15.3 months the patients received rituximab (RTX) total dose of 2.5±1.3 grams in combination with glucocorticoids at average dose of 11.7±3.9 mg. 10 (24%) patients concurrently took immunosuppressants. The therapy efficacy was evaluated both in the general study population and in the patient subgroups with interstitial lesion extent up to 20% (Group A, n=13) and greater than 20% (Group B, n=29) of total pulmonary tissue area.¹

Results: In the general patient population significant FVC increase from 73.2±18.8% to 82±21.8% (p=0.000031) and stabilisation of DLCO (42.6%±15.7% vs 44.7±14.6%. p=0.2) were observed. Median FVC increment was 6% (25th% >3.3%; 75th%>16%). FVC-based parameters increased by ≥10% in 16 (38%) patients and decreased in 3 (7%) patients.

Average FVC values in Group A were significantly higher compared with Group B both at the baseline (88.8%±18.6% vs 65.4±14.5%, p=0.0002) and after the treatment (103.3%±15.9% vs 74.1±18.5%, p=0.0009) with statistically significant FVC increase in both groups during the treatment period (p=0.016 and p=0.0014, respectively). Median FVC increment in Group A and Group B was 10.2% (25th% >4.7%; 75th%>21.9%) and 5.9% (25th%>2.75%; 75th%>14.7%), p>0.05, respectively. FVC-based parameters increased by ≥10% in 6 (46%) patients in Group A, and in 10 (34%) patients in Group B, and decreased in 1 (8%) and 2 (7%) patients, respectively.

Average DLCO values were also significantly higher in Group A compared to Group B both before and after treatment (58.4%±16.4% vs 36.3±10.1%. p=0.025; 59.3±15.2% vs 38.9±9.7%. p=0.005); DLCO values did not change over time during RTX therapy.

Conclusions: RTX therapy resulted in significant FVC increase. FVC increment in the patient group with ILD extent up to 20% achieved clinical significance level in contrast to the patients with ILD extent greater than 20%, where FVC increment was 5.9%. Obtained data suggest that initial lung lesion area is a potential predictor of response to a-B-cell therapy in the patients with SSc.

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THU0421

PREVALENCE AND CLINICAL CORRELATES OF SMALL AIRWAY OBSTRUCTION IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Small airways are usually defined as non-cartilaginous airways with an internal diameter <2 mm. Small airway obstruction (SAO) may be result of a primary bronchiolar disorder, or secondary to a disease that also affects large airways (like asthma or chronic obstructive pulmonary disease - COPD), or related to an interstitial lung disease with bronchiolar involvement.

Objectives: To assess prevalence and clinical correlates of SAO in patients with systemic sclerosis (SSc).

Methods: 69 consecutive patients with SSc (63 women and 6 men) were included in this study. Patients with previously diagnosed bronchiectasis, COPD or asthma were excluded. Forty two (60.9%) patients had limited cutaneous SSc, whilst 27 (39.1%) of patients had diffuse form of the disease. Seventeen patients (24.6%) were tobacco-smokers, 52 (75.4%) were nonsmokers. Lung function tests, including assessment of lung diffusing capacity, were performed in all patients. Patients were considered to have small airway obstruction (SAO) when Maximum Expiratory Flow at 25% of the forced vital capacity (MEF₂₅) was lower than 60% as predicted. We assessed the relationship of SAO in our patients with large airway obstruction, decreased lung diffusing capacity, disease duration, disease subtype, scleroderma-specific antibodies and smoking.

Results: SAO was noticed in 46/69 (66.6%) of patients with SSc. Restrictive lung disease was found in 4/69 (5.8%), obstruction of large airways in 18/69 (26.1%) and decreased lung diffusing capacity in 47/69 (68.1%) of patients. No difference in gender, age, disease duration, disease form and scleroderma-specific antibodies was found between patients with and without SAO. 18/46 (39.1%) patients with SAO had decreased FEV₁ and FEV₁/FVC, indicating presence of coexistent large airway obstruction. Indeed, all patients with signs of obstructive lung disease on spirometry, had associated SAO. Moreover, MEF₂₅ correlated significantly with FEV₁ (ρ=0.54, p<0.001), FEV₁/FVC (ρ=0.74, p<0.001), PEF (ρ=0.29, p=0.02) and MEF₅₀ (ρ=0.80, p<0.001) in our patients with SSc. However, 28/46 (60.9%) SSc patients with SAO did not have signs of concomitant large airway obstruction. 20/46 (43.5%) patients had associated decreased lung diffusing capacity, whilst 8/46 (17.4%) of patients had isolated SAO. We have found that tobacco-smokers among SSc patients with SAO have more common associated obstructive lung disease on spirometry (58.8% vs 15.4%, p=0.004), and have a significantly lower mean value of MEF₂₅ compared to non-smokers (32.1%±7.1% vs. 45.4±11.6%, p=0.001). On the other hand, isolated SAO, and SAO associated with impaired diffusing capacity was equally frequent among smokers and non-smokers.

Conclusions: Patients with SSc have commonly SAO. It can be considered as clinical feature of undiagnosed asthma or COPD, if associated with large airway obstruction, especially in tobacco-smokers. On the other hand, isolated SAO or associated with decreased lung diffusing capacity was found to be not related to smoking, and may indicate a possible prominent bronchiolar involvement within SSc related interstitial lung disease.

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THU0422

PERFORMANCE OF EULAR/ACR 2017 IDIOPATHIC INFLAMMATORY MYOPATHIES CLASIFICACION CRITERIA IN A REAL WORLD COHORT

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Background: Idiopathic Inflammatory Myopathies (IIM) are a heterogeneous group of multisystemic diseases. It includes Polymyositis (PM), Dermatomyositis (DM) with its clinically amyopathic variant (CADM), the antisynthetase syndrome (ASS), the inclusion body myositis (IBM), Immune-mediated necrotising myopathy (IMNM), the juvenile variants of DM/PM and the connective tissue disease-myositis overlap (CTD-OM).

Distinction between subtypes is made on grounds of clinical features, histologic findings at muscle biopsy and presence of certain autoantibodies.

Multiple classification criteria had been proposed through time, EULAR/ACR been the most recent. However, their performance in patients from common practice in Latin America had not been widely evaluated.

In our practice, access to muscle biopsy and electromyogram (EMG) is not always available

Objectives: To evaluate the performance of the EULAR/ACR 2017 IIM classification criteria in a real world cohort and compare it with the performance of other classification criteria

Methods: Retrospective study. IIM patients defined by expert opinion followed in our centre between October 2007 and November 2017 were included.

The patients were classified clinically in DM, CADM, PM, ASS and CTD-OM. Patients with positive antisynthetase antibodies were reclassified as ASS.

Availability of EMG, muscle biopsy and anti Jo-1 antibodies was evaluated. , Bohan & Peter (1975) Tanimoto (1995) y EULAR/ACR (2017) criteria were applied to the population

Results: 60 patients were included. DM 20 (33.3%), CADM 4 (6.66%), PM 4 (6.66%), ASS 10 (16.66%) y CTD-OM 22 (36.66%). Muscle biopsy available 14/60 (23.33%), EMG available 33/60 (55%) and anti Jo-1 determination available in 57/60 (95%).

In general, 12/60 (20%) classified as defined disease by Bohan and Peter criteria, 29/60 (48.3%) by Tanimoto criteria and 34/60 (56.6%) by EULAR/ACR 2017 criteria.