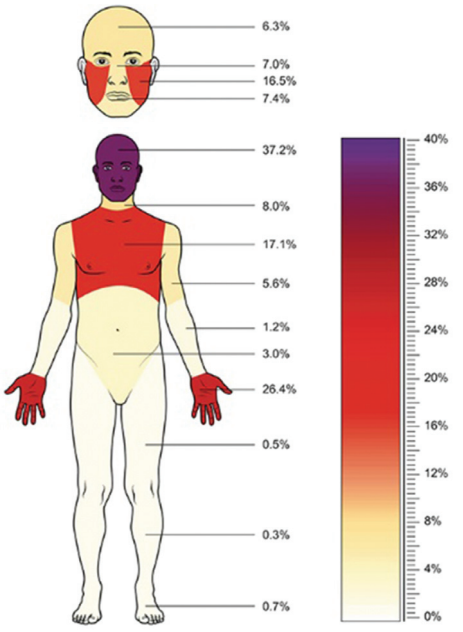

Conclusions: In SSc-patients, TA were predominantly located on the face, hands and the upper part of the trunk. They may reflect the vasculopathy of SSc and could represent a clinical biomarker for vascular disease, particularly for PH, one of the most severe vascular complications of the disease.

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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)

M. Pestaña-Fernández[1], M. Rubio-Rivas[1], N. Villalba-Jiménez[1], B. Marí-Alfonso[2], A. Guillén-Del-Castillo[3], M. Freire[4], J.A. Vargas-Hitos[5], J.A. Todolí-Parra[6], M. Rodríguez-Carballeira[7], G. Espinosa-Garriga[8], X. Corbella Virós[1], V. Fonollosa-Pla[3], C.P. Simeón-Aznar[3] on behalf of RESCLE. *[1]Internal Medicine, Hospital Universitario Bellvitge, Barcelona*; *[2]Internal Medicine, Corporació Sanitària Parc Taulí, Sabadell*; *[3]Internal Medicine, Hospital Universitario Vall d'Hebron, Barcelona*; *[4]Internal Medicine, Complejo Hospitalario Universitario de Vigo, Vigo*; *[5]Internal Medicine, Hospital Universitario Virgen de las Nieves, Granada*; *[6]Internal Medicine, Hospital Universitario y Politécnico La Fe, Valencia*; *[7]Internal Medicine, Hospital Universitario Mútua Terrassa, Terrassa*; *[8]Internal Medicine, Instituto Clínic de Medicina y Dermatología. Hospital Universitario Clínic, Barcelona, Spain*

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-mortaliy 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 endpoint 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 freqüent 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 wtih moderate ILD.
Upfront combination therapy may improve survival, but did not reach statistical significants 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 RATEWITH PULMONARY FUNCTION TESTS AND EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (ESCSG-AI) IN SYSTEMIC SCLEROSIS – ASSOCIATED INTERSTITIAL LUNG DISEASE IN FOLLOW UP STUDY

O.B. Ovsyannikova, O. Koneva, L. Ananieva. *Department Of Vascular Pathology, Nasonova Research Institute of Rheumatology, Moscow, Russian Federation, Москва, Russian Federation*

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=0309 (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.