± 0.2 vs 0.3 ± 0.2). Fifteen/20 (75%) had an abnormal ($<90\%$ within 10 sec) esophageal emptying activity. Percentage of esophageal emptying activity negatively correlated with reflux score ($r = -0.52$, $p = 0.02$) while it did not correlate with the other items scales and the total UCLA SCTC_GIT 2.0 score ($p > 0.05$).

Conclusion: SSc patients with impaired esophageal scintigraphy findings have a higher GIT2 reflux score. The lack of correlation with UCLA SCTC_GIT 2.0 total score may be related to the composite nature of the score capturing overall GI disease aspects. The UCLA SCTC_GIT 2.0 is a complementary tool for objective measurement of esophageal involvement which can be easily administered in day-to-day clinical assessment. A larger number of SSc patients is needed to confirm these preliminary findings.

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FRI0311

PREVALENCE AND CLINICAL MANIFESTATIONS OF ERASMUS SYNDROME IN SYSTEMIC SCLEROSIS: A CROSS-SECTIONAL STUDY

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Background: Erasmus syndrome (ErS) is defined by the association of exposure to silica with the subsequent development of systemic sclerosis (SSc), with or without associated silicosis.

Objectives: The objectives of this study were, on one hand, to evaluate the prevalence of ErS in a population of SSc patients and to characterize the cases and, on the other hand, to evaluate the clinical and laboratory characteristics of SSc patients with or without exposure to silica.

Methods: We performed a cross-sectional study of the patients with SSc in our department. Demographics, clinical and laboratory data were collected from all patients with SSc diagnosed according to ACR/EULAR criteria. Moreover, a telephone call was made in order to detail the professional activity and possible exposure to silica.

Results: The prevalence of ErS in this population was 15.3% (9/59). All cases identified were male, corresponding to 75% of men with SSc followed at our department. There was a statistically significant association between ErS and male gender ($p < 0.001$), initial pulmonary manifestation ($p = 0.025$), history of digital ulcers ($p = 0.014$) and smoking ($p = 0.047$). On the other hand, a lower risk of gastrointestinal involvement was found in ErS cases ($p = 0.008$). All patients with ErS had positive autoantibodies (mainly anti-Scl70 and anti-centromere) with titers tending to be higher than SSc without ErS, although without statistically significant differences. In addition, although with no statistical significance, we found that pulmonary artery systolic pressure (PASP) estimated by echocardiogram was higher in patients with ErS.

Conclusion: In our study, prevalence of ErS was higher than data from previously published literature. For a more accurate ErS diagnosis it is necessary to be aware of and investigate less intense silica exposures, which may have occurred many years before diagnosis. Statistically significant differences were found between ErS and SSc without exposure to silica; this fact may have impact in diagnosis, treatment and prognosis.

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FRI0312

HYDROXYCHLOROQUINE SIGNIFICANTLY REDUCES SERUM MARKERS OF ENDOTHELIAL INJURY AND NEMO VIDEOCAPILLAROSCOPY SCORE IN SYSTEMIC SCLEROSIS: A 3-MONTHS PROSPECTIVE OBSERVATIONAL STUDY

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Background: The most commonly postulated model of disease progression in Systemic sclerosis (SSc) is based on the interplay between endothelial damage and tissue repair. Hydroxychloroquine (HCQ) is commonly used in systemic sclerosis (SSc) for the management of joint involvement [1]. In addition, HCQ showed to exert a protective role on endothelial dysfunction in several pathologic conditions [2] but no data exist in SSc.

Objectives: aim of our study was to assess the effect of 3-months HCQ administration on serum levels of markers of endothelial injury and microvascular changes, assessed with NEMO score.

Methods: 20 SSc patients considered for treatment with HCQ 400 mg/day and 16 SSc patients not treated with HCQ were consecutively recruited, having clinical assessment, blood samples and nailfold capillaroscopy (NVC) at recruitment and after 3 months. Median age was 51.5 (19.5). Median onset of Raynaud's phenomenon was 83 (66) months. 29 patients had ISSc and 7 patients dSSc. Both groups had comparable vasoactive drugs, disease activity, duration and subtype. Any kind of overt vasculopathy was an exclusion criteria. The NEMO score was calculated as the total number of micro-haemorrhages (MHE) and microthrombosis (MT) and the giant capillaries (GC) score was calculated as the total number of GC [3]. Serum E-selectin, Endothelin-1 (ET-1), Vascular cell adhesion molecule-1 (VCAM-1), vascular endothelial growth factor (VEGF-A) levels were assayed by Magnetic Luminex Assay, Human Premixed Multi-Analyte Assay (R&D Systems). Plasma dimethyl arginine (ADMA) levels were determined by competitive ELISA (DLD Diagnostica GmbH). Data were assessed with SPSS software version 25.0 and expressed as median and interquartile range. Wilcoxon rank-sum test, Mann-Whitney U test and Fisher's exact test were used for statistical evaluation. P values < 0.05 were considered significant.

Results: in the group treated with HCQ serum levels of E-Selectin (35.28 (16.88) ng/ml vs 30.84 (12.60) $p = 0.004$), VCAM-1 (847.30 (344.85) ng/ml vs 761.55 (396.87) $p = 0.014$) and ET-1 (12.25 (5.53) pg/ml vs 10.94 (4.35) pg/ml ($p = 0.022$) decreased significantly, as well as NEMO Score (7.5 (11.25) vs 6.5 (8) $p = 0.001$), MHEs (6 (7) to 5 (5.75) $p = 0.003$) and MTs (1 (4.5) to 0.5 (2) ($p = 0.003$)). VEGF-A ($p = 0.68$), ADMA ($p = 0.91$) and GCs ($p = 0.30$) were unaffected. No significant improvements were observed among patients not treated with HCQ.

Conclusion: a significant improvement in endothelial function and microvascular involvement was achieved after three months of HCQ treatment. It is a novel finding in SSc which can represent a new therapeutic possibility for the prevention of microvascular complications of the disease

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FRI0313 DIFFERENTIAL PERFORMANCE OF NAILFOLD VIDEO CAPILLAROSCOPIC PARAMETERS IN THE DIAGNOSIS AND PROGNOSIS OF SYSTEMIC SCLEROSIS

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Background: The role of Nailfold Video Capillaroscopy (NVC) in the identification of patients with Raynaud phenomenon (RP) at risk to develop systemic sclerosis (SSc) is well established. However, it is not clear if certain capillaroscopic indices perform better than others at predicting SSc development and which NVC parameters have a prognostic value in established SSc.

Objectives: To comparatively assess the performance of different NVC parameters in predicting development of SSc, very early diagnosis of SSc (VEDOSS), or mixed connective tissue disease (MCTD) in patients with RP. Also, to longitudinally examine the consistency of clinical correlations of NVC parameters in SSc patients at two different time points and evaluate the prognostic capacity of NVC in SSc.

Methods: Consecutive RP patients referred to our department for NVC (138 with SSc, 12 with VEDOSS, 6 with MCTD, 36 with primary RP, and 50 with non-SSc secondary RP) were evaluated at baseline, both clinically and capillaroscopically; 175 were reevaluated after a mean±SD of 3.34±1.48 years. Sixty-two healthy volunteers served as controls. Qualitative assessment of NVC images permitted categorization of patients to a normal, early, active or late capillaroscopic pattern. Capillary loss, dilated, giant or ramified capillaries and micro-hemorrhages were evaluated by a semi-quantitative score (0-3), derived as the mean of three 1mm fields in each of the 2nd, 3rd, 4th and 5th finger of both hands. FVC and DLCO were recorded, if performed within 6 months of NVC. FVC and DLCO deterioration were considered clinically significant if >10% and >15%, respectively. Skin thickening was measured using the modified Rodnan Skin Score (mRSS). mRSS deterioration was considered clinically significant if >3.5. First occurrence of digital ulcers in patients with no prior such history and vital status were also recorded at follow-up.

Results: Capillary loss score had the highest diagnostic accuracy at discriminating patients with an SSc-spectrum disorder from patients with RP of different etiology and from controls, as defined by ROC curve analysis [AUC (95% CI)=0.925 (0.893-0.956)], followed by dilatation score [AUC (95% CI)=0.904 (0.807- 0.938)] and giant score [AUC (95% CI)=0.856 (0.810-0.902)]. By contrast, micro-hemorrhages [AUC (95% CI)=0.727

(0.669-0.786)] and ramification scores [AUC (95% CI)=0.588 (0.521-0.654)] did not perform equally well. Notably, clinical correlations of capillaroscopic parameters in SSc were found not to be consistent over time, when longitudinally assessed at two different time points by univariate and multivariate analysis. Binary logistic regression analysis indicated that baseline capillaroscopic pattern could predict occurrence of a combined adverse disease outcome (FVC deterioration>10% and/or DLCO deterioration>15% and/or mRSS deterioration>3.5 and/or first occurrence of digital ulcers and/or death), after a mean±SD follow up of 3.28±1.45 years in 94 SSc patients with available follow-up data (OR=3.43, p=0.031 for active versus early pattern, OR=8.778, p=0.007 for late versus early pattern).

Conclusion: Dilatation score performs best of all semi-quantitative NVC parameters in diagnosing SSc and although clinical correlations of capillaroscopic findings change over time, an active or late capillaroscopy pattern at baseline is associated with an adverse prognosis.

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FRI0314 SENSITIVITY TO CHANGE AND RESPONSIVENESS TO TREATMENT OF RENAL RESISTIVE INDEX (RRI) IN SYSTEMIC SCLEROSIS (SSC)

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Background: RRI may identify any problem with the blood flow in the renal artery and in the parenchyma. It was shown to be increased in SSc patients and being in relationship with both vascular and fibrotic SSc-related manifestations.

Objectives: to test sensitivity to change and responsiveness to treatment of RRI in SSc.

Methods: patients fulfilling the 2013 ACR/EULAR classification criteria for SSc were enrolled from two SSc-care units, if RRI was determined at least twice since being diagnosed. Data regarding SSc clinical manifestations, instrumental and laboratory evaluation for renal, cardiac and cardiovascular involvement, as well as ongoing immunosuppressive and vasoactive/vasodilating treatment, were collected both at baseline and follow-up RRI measurements.

Results: 230 patients [aged 57 (48-67) years, 12.6% males] were enrolled in the study, with baseline RRI value of 0.68 (IQR 0.07). In a mean 3.4 years follow-up, RRI showed a median change (ΔRRI) of +0.02 (IQR 0.05). ΔRRI was significantly correlated with ΔsPAP (R=0.173, p=0.023) and it was significantly higher in patient with new onset of pulmonary arterial hypertension (0.08 ± 0.02 vs 0.03 ± 0.05; p=0.038). On Cox univariate regression analysis, time from disease onset, ΔssPAP and ΔRRI predicted new PAH, while ΔsPAP was the only independent predictors at multivariate regression analysis. Regarding treatment, Sildenafil exposure determined a significantly lower increase in ΔRRI, with a progressive long-term effect. Conversely, CCBs and Iloprost treated patients showed a significantly higher increase of ΔRRI, which was determined by a higher DU burden. No difference was seen when immunosuppressive treatment was evaluated.

Conclusion: RRI is sensitive to change and reflects, in particular, the worsening of cardio-pulmonary vascular involvement (increase in ΔsPAP and new PAH onset). Moreover, it shows a possible protective effect of Sildenafil in reducing pulmonary vasculature manifestations.

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