test 8 (MT8) were collected. Data were reported as median interquartile range; non-parametric tests were used for the analysis.

**Results:** Subjects admitted presented muscular edema in at least one muscle. Patients with muscular edema had higher CK levels (1506±1976 vs 235±224 p<0.001) lower disease duration (9.5±47.0 vs 19.8±97.2 p=0.031). and lower MT8 (63.6±11.2 vs 69.8±10.7 p=0.03). Forty-eight patients presented fatty infiltrates that were more frequent in older patients (63.1±11-3 vs 52.8±13.7 p=0.01) and in those with longer disease duration (39.1±95.4 vs 9.5±12.5 p=0.024). CK levels and MT8 were not different in patients with or without muscular fatty infiltrates. With the multivariate analysis the disease duration represents the only independent factor for the presence of muscular fatty infiltrates (p=0.05).

**Molecular atrophy was present in 17 patients but it was not correlated to age and disease duration.** CK and MT8 were not different in presence/absence of fatty infiltrates or muscular atrophy.

**Edema and atrophy were not different between poly- and dermatomyositis.** The fatty infiltrate was more present in the posterior compartment of the tights (biceps femoris and semitendinosus muscle) in patients with PM compared to DM (68.2% vs 29.6% p=0.046).

**Conclusion:** The alterations identified with MRI in our cohort changed according to the disease duration and the age of the patients. In particular, fatty infiltration was more frequent in patients with longer disease duration, but was not associated to CK levels and muscular weakness. Moreover, fatty infiltration was prevalent in the posterior compartment of the tights in PM patients.

**Molecular MRI is widely used in the diagnosis and follow-up but the clinical meanings of the different alterations still need to be investigated.**

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**SA0261**

**CHARACTERISTICS ASSOCIATED TO SCLERODERMAL RENAL CRISIS, AND INCIDENCE VARIATION OVER TIME IN THE RESCLE REGISTRY**

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**Background:** Scleroderma Renal Crisis (SRC) is a serious complication of Systemic Sclerosis (SSc). Nowadays, it seems that there is a reduction in its prevalence and mortality1.

**Objectives:** To evaluate the characteristics of patients with SRC in a large cohort of SSc patients. To investigate predictors of SRC, and epidemiologic differences over time.

**Methods:** 1933 patients were collected in ongoing registry of Spanish SSc patients – RESCLE. We did descriptive study and epidemiologic analysis.

**Results:** Out of 1933 SSc, 43 (2.2%) developed SRC. Univariate analysis showed significant differences of SRC vs. non-SRC cases: SSc subtypes: diffuse cutaneous SSc (dcSSc), 72% vs. 19%; limited cutaneous SSc (lcSSc), 26% vs. 61%. Demographics: Female gender, 77% vs. 89%; time from SSC onset to SSC diagnosis, 3.0±8.0 vs. 6.6±9.5 years; arterial hypertension (HT), 56% vs. 32%. 1st manifestation: Raynaud’s phenomenon (RP), 68% vs. 82%. Clinical manifestations: RP, 86% vs. 96%; digital ulcers, 70% vs. 38%; arthritis, 45% vs. 20%; myositis, 30% vs. 13%; joint contractures, 45% vs. 18%; intestinal involvement, 24% vs. 11%; interstitial lung disease 58% vs. 41%; pulmonary HT, 56% vs. 29%; pericardial effusion, 28% vs. 7.4%; pericarditis, 24% vs. 8.3%; ischemic cardiopathy, 31% vs. 12%; diastolic dysfunction, 67% vs. 34%. Capillaroscopy: active pattern 77% vs. 33%. Immunological data: anti-Topoisomerase I, 39% vs. 20%; anti-centromere, 15% vs. 49%; anti-RNAPol III, 45% vs. 11%. Prognosis: Overall mortality, 56% vs. 18%; SSc-related mortality, 83% vs. 49%. Survival at 5, 10, 20, and 30 years was 73% vs 98%, 56% vs. 92% and 51% vs. 25%. Multivariate analysis: dcSSc subtype, RR 22.68 (5.81-88.51) p<0.001; intestinal malabsorption, RR 7.35 (2.55-21.18) p<0.001, and active capillaroscopy pattern RR 7.26 (1.61-32.7) p<0.010. Prevalence of SRC (P) in dcSSc subtype 7.8%, and in lcSSc subtype 0.9%. P over decades: P<0.1, 3.8%; P<0.10, 2.7%; P<0.01, 2.3% and P<0.01, 0.9% achieving statistical significance in the last decade RR: 0.33 (0.13-0.85) p=0.014.

**Conclusion:** In RESCLE cohort, SRC predominated in dcSSc patients, and it was associated to intestinal malabsorption, and an active pattern by capillaroscopy. SRC showed a very poor prognosis. Finally, we evidenced a decreasing prevalence of SRC over time in our cohort.

**REFERENCE**


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**SA0262**

**MODIFIED ACR COMPOSITE RESPONSE INDEX IN SYSTEMIC SCLEROSIS SCORE SHOWS SENSITIVITY AND EXTERNAL VALIDATION TO MEASURE MAGNITUDE OF RESPONSE AT 12 MONTHS IN DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS**

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**Background:** The Composite Response Index in Systemic Sclerosis (CRiSS) is a 2-step process for the probability of improvement of patients with diffuse cutaneous systemic sclerosis (dcSSc) ranging from 0.0 (no improvement) to 1.0 (1). The limitation of probability is that it does not measure magnitude of response and it cannot be negative, hence reflecting an improvement.

**Objectives:** Here we aimed to test the performance of CRiSS numerator normalized by baseline values to assess% change at 12 months in an observational cohort.

**Methods:** Consecutive dcSSc patients were included in the study at a single centre. Clinical data were collected at baseline and 12 months and CRiSS calculated using the published formula. To explore the quantitative value of the numerator, each of the 5 domains was corrected for their baseline value (e.g (12 months baseline)/baseline = Delta%) while keeping their original relative weight in the formula. The weighted sum was then divided by the number of domains. The modified CRiSS (mCRiSS) was then compared with the original CRiSS score and with another composite score calculated on detrended data. The Global Ranked Composite Score (GRCS) (2) was used in treating organ failure to assign the worst negative values. Spearman’s test was used for Prism 7 correlation analysis. P < 0.05 was considered significant.

**Results:** Thirty-three dcSSc patients were enrolled. Twenty-one patients had a CRiSS < 0.0%. Twelve patients had a positive CRiSS ranging from 1% to 77%. 18/21 patients with CRiSS > 0 showed a negative mCRiSS (negative response = worsening) ranging from -9% to -2500%. 10/12 with positive CRiSS showed a corresponding positive mCRiSS ranging from 1% to 22%. mCRiS had a significant correlation with CRiSS (r = 0.5, p = 0.003) and GRCS (r = 0.7, p < 0.0001) despite the low number of patients enrolled.

**Conclusion:** Normalization by baseline data within CRiSS numerator offers a quantitative score to assess magnitude of response, considering the 5 domains and their relative weight within the original CRiSS. The mCRS