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 mortality. The objective of this study was to evaluate the characteristics of SSc patients with SRC in a large cohort of SSC patients. To investigate predictors of SRC, and epidemiologic differences over time.

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

RESULTS: Out of 1933 SSc patients, 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±2.0 vs. 6.6±9.5 years; arterial hypertension (HT), 56% vs. 32%; 1st manifestation: Raynaud’s phenomenon (RP), 68% vs. 82%; Clinical manifestations: RP, 68% 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%; malabsorption, 24% vs. 9%; idiopathic 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-TPOsierosimerase I, 39% vs. 20%; anti-centromere, 15% vs. 49%; anti-SSA/SSB, 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. 96%, 56% vs. 92%, 53% vs. 92%, 38% vs. 67%, respectively. Treatment: ACEI use, 35% vs. 14%, corticoid use, 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 (F) in dcSSc subtype 7.8%, and in lcSSc subtype 0.9%. P over decades: P<0.05, 80% vs. 90%, P<0.05, 2.7%, P<0.05, 2.3% and P<0.05, achieving statistical significance in the last decade RR: 0.33 (0.13-0.85) p<0.014.

CONCLUSION: SRC predominantly 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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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 expected worsening.

OBJECTIVES: Here we aimed to test the performance of CRiSS numerator normalised 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 validated in randomized controlled trials, the Global Ranked Composite Score (GRCS) (2). Death and organ failure were also included and assigned 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%. mCRiSS 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 mCRiSS