years; 92% were women, and the average time of illness was 8.2 years. The model based on theory showed that age (aPR = 1.03, 95% CI = 0.99-1.07, $p = 0.143$) and exposure to Penicillamine (aPR = 0.51, CI 95% = 0.19-1.33, $p = 0.170$) were marginally associated ($p < 0.2$) with CKD, while pulmonary hypertension (aPR = 2.76, 95% CI = 1.29-5.89, $p = 0.009$) and arterial hypertension (aPR = 3.51, 95% CI = 1.06-11.6, $p = 0.04$) were significantly associated (< 0.05) with CKD. The parsimonious model retained pulmonary hypertension (aPR = 3.74, 95% CI = 1.67-8.36, $p = 0.001$) and arterial hypertension (aPR = 7.45, 95% CI = 3.31-16.7, $p < 0.001$) as significantly associated factors to CKD.

Conclusion: Hypertension, a classic cardiovascular risk factor, and pulmonary hypertension were important factors associated with CKD. The appropriate management of these factors must be taken into account to prevent CKD. Prospective cohort studies should evaluate the influence of these factors in reducing the incidence of CKD.

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AB0214 MUSCLE INVOLVEMENT IN SYSTEMIC SCLEROSIS

Boris Kisluk¹, Jose Milisenda^{2,3}, Iago Pinal-Fernandez⁴, Pedro Juan Moreno Lozano², Alfredo Guillén-Del-Castillo⁵, Sergio Prieto-González², Gerard Espinosa², Carmen Pilar Simeón-Aznar⁵, Albert Selva-O'callaghan⁵, Josep Maria Grau^{2,3}.
¹Hospital Escuela Eva Perón, Rosario, Argentina; ²Hospital Clínic de Barcelona, Barcelona, Spain; ³CIBERER, Barcelona, Spain; ⁴National Institutes of Health, National Institute of Arthritis and Musculoskeletal and Skin Diseases, Bethesda, United States of America; ⁵Vall d'Hebron General Hospital, Systemic Autoimmune Diseases Unit, Barcelona, Spain

Background: The prevalence of myopathy in systemic sclerosis (SSc) or scleroderma patients varies from 5% to 81% depending on the diagnostic criteria used to define the muscle involvement. The histopathological characteristics found in these patients and their correlation with the clinical features and autoantibody profile has not been fully characterized.

Objectives: To characterize the muscle involvement and histopathology findings in patients with SSc.

Methods: This retrospective cross-sectional study included all patients from Scleroderma Cohort of Vall d'Hebron General and Hospital Clínic de Barcelona with muscle biopsies available for review performed at the Muscle Research Unit of Hospital Clínic de Barcelona from May 2004 to November 2018. Muscle biopsies were performed, for weakness or raised CK and/or aldolase. Histological sections were independently evaluated by two myopathology experts looking for inflammation in the endomysium, perimysium and perivascular areas. The presence of necrosis, regeneration, fibrosis, neurogenic atrophy, and fiber type grouping as well as perifascicular atrophy were also recorded. Based on the autoantibody profiles patients were classified into five groups, "Scl70", "PM/Scl", "centromere", "U1-RNP", and "nucleolar and centromer IFI ANA pattern without specificity for anti-Scl70 or anticentromer". Demographic and clinical data were obtained by chart review.

Results: 42 subjects were included, 33 (78.6%) of them were women. Considering the immunological profile 12 had Scl70, 12 PM/Scl, 5 anticentromere, 2 U1RNP, and 11 had ANA pattern without specificity for anti-Scl70 or anticentromer. Most of patients (90.5%) were Caucasians, 2 (4.8%) were Hispanic, and 2 of other ethnic background. The mean age at the onset of SSc was 42.1 (SD 2.5) years and at the muscular symptoms 50.7 (SD 2.1) years, respectively. Twenty-two (52.4%) patients presented with high CK level (mean level of 1316 ± 353.8 U/L; normal value ≥ 200 U/L) whereas 38 (90.5%) exhibited high aldolase level (mean 19 ± 2.53 U/L, normal value < 6 U/L). Of note, 17 (40.5%) presented with high aldolase level and normal CK. Regarding the clinical manifestations of muscle involvement, 34 (80%) patients developed symmetrical distribution. Twenty-six (61.9%) had interstitial lung disease and 5 (11.9%) patients developed cancer.

Among the pathological features, 28 (66.7%) biopsies had fibrosis. Neurogenic atrophy was detected in 4 (9.5%) and fiber type grouping was present in 9 (21.4%) muscle biopsies. In near half of the muscle biopsies (45.2%) inflammatory infiltrate was present. Compared to other patients, those positive for PM/Scl had more perivascular (75%),

endomysial (50%) and perimysial inflammation (75%) and perifascicular atrophy (42%) (all $p < 0.05$), while a reduced prevalence of inflammatory infiltrates was noted in the Scl70 group (17%) ($p < 0.05$).

Conclusion: In our cohort of SSc patients with muscle involvement, two main histopathological patterns were found, fibrosis and inflammation. Almost half patients presented with elevated aldolase with normal CK levels. The muscle disease heterogeneity suggests that a variety of pathologic mechanisms play a role in the scleroderma associated myopathy but those patients with histopathological inflammatory features deserve to be treated with immunosuppressive therapy.

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AB0215 A LATE ONSET OF SYSTEMIC SCLEROSIS IS ASSOCIATED WITH A MORE RAPIDLY PROGRESSING CLINICAL PHENOTYPE IN LCSSC PATIENTS

Pia Moizadeh¹, Kathrin Kuhr², Ulf Müller-Ladner³, Elise Siegert⁴, Jörg Henes⁵, Gabriela Riemekasten⁶, Norbert Blank⁷, Marc Schmalzing⁸, Ina Koetter⁹, Claudia Guenther¹⁰, Tim Schmeiser¹¹, Gabriele Zeidler¹², Alexander Kreuter¹³, Margitta Worm¹⁴, Laura Susok¹⁵, Aaron Juche¹⁶, Christiane Pfeiffer¹⁷, Cord Sunderkoetter¹⁸, Donja Homayoon¹⁹, Nicolas Hunzelmann¹.
¹University Hospital Cologne, Dermatology and Venerology, Cologne, Germany; ²University of Cologne, IMSB, Cologne, Germany; ³Justus Liebig University Giessen, Campus Kerckhoff, Rheumatology, Bad Nauheim, Germany; ⁴Charité – Universitätsmedizin Berlin, Rheumatology, Berlin, Germany; ⁵Center for Interdisciplinary Clinical Immunology, Rheumatology and Autoinflammatory Diseases, University Hospital Tuebingen, Rheumatology, Tuebingen, Germany; ⁶University Medical Center - UKSH, Rheumatology, Luebeck, Germany; ⁷University Hospital Heidelberg, Rheumatology, Heidelberg, Germany; ⁸University Hospital Würzburg, Rheumatology, Würzburg, Germany; ⁹Asklepios Clinic Altona, Rheumatology, Hamburg, Germany; ¹⁰University Hospital Carl Gustav Carus Dresden, Dermatology, Dresden, Germany; ¹¹Hospital St. Joseph, Rheumatology, Wuppertal, Germany; ¹²Johanniter-Krankenhaus im Fläming Treuenbrietzen, Rheumatology, Treuenbrietzen, Germany; ¹³HELIOS St. Elisabeth Hospital Oberhausen, University Witten-Herdecke, Dermatology, Oberhausen, Germany; ¹⁴Charité – Universitätsmedizin Berlin, Dermatology, Berlin, Germany; ¹⁵Ruhr-University Bochum, St. Josef Hospital, Dermatology, Bochum, Germany; ¹⁶Immanuel Hospital Berlin, Rheumatology, Berlin, Germany; ¹⁷University Hospital Ulm, Dermatology, Ulm, Germany; ¹⁸University Hospital Halle (Saale), Dermatology, Halle, Germany; ¹⁹Medical University of Graz, Dermatology, Graz, Austria

Background: SSc is a heterogeneous multisystem connective tissue disease. The majority of patients (pts) develop initial clinical symptoms between the ages of 30-50 years (yrs).

It is not yet known whether the age of onset has an influence on the development of a distinct clinical phenotype.

Objectives: Investigating the relationship between age at disease onset and the development of clinical characteristics using data of the German Network for Systemic Scleroderma.

Methods: We evaluated 2928 patient data, subdivided them into 3 age groups at disease onset (<40 years, 40-60 years, and >60 years) and correlated the age at disease onset with specific clinical characteristics.

Results: Overall, 24% of pts developed first non-RP symptoms at the age of <40yrs, 53% between the age of 40-60yrs, and 23% were older than 60yrs. In particular, SSc pts with disease onset >60yrs developed significantly ($p < 0.001$) more often the lcSSc subtype (64%) as well as ACA antibodies (42%) with a significantly lower mRSS. They, however, also suffered more often from pulmonary hypertension (PH) and developed less often digital ulcers. Especially lcSSc pts were associated with a more rapidly progressing clinical phenotype, i.e., 25% of the elderly lcSSc pts developed PH after five years. In contrast, less than 5% diagnosed with lcSSc at the age of <40yrs suffered from PH after five yrs ($p < 0.001$).

Conclusion: In this registry, approximately one quarter of pts developed SSc at an age above 60yrs, predominantly having a lcSSc subtype. Although these pts have been diagnosed with the mild form of SSc, pts with a lcSSc subtype at a higher age (>60yrs) had more frequent a PH and showed a more rapid disease progression than the youngest pts. These findings may have an important influence on recommendations on