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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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.8192

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 anti-centromere, 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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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.6007

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

diagnosis, frequency of follow-ups and therapy of SSc in different age groups.

Disclosure of Interests: Pia Moinzadeh Speakers bureau: Actelion, Kathrin Kuhr: None declared, Ulf Müller-Ladner Grant/research support from: Projekt supported by an unrestricted educational grant from Celgene GmbH., Elise Siegert Shareholder of: Astra Zeneca, Grant/research support from: Actelion, Consultant for: AEC Partners, Speakers bureau: Actelion, Norsk, Jörg Henes: None declared, Gabriela Riemekasten Consultant for: Chugai, F. Hoffmann-La Roche, Speakers bureau: Chugai, F. Hoffmann-La Roche, Norbert Blank Grant/research support from: SOBI and Novartis, Speakers bureau: Novartis and SOBI, Marc Schmalzing Grant/research support from: Pfizer, Chugai, MSD, Janssen-Cilag, BMS, Celgene, UCB, Consultant for: Abbvie, Chugai, Genzyme, Hexal/Sandoz, MSD, Novartis, Roche, Sanofi Pasteur, Speakers bureau: Actelion, Baxalta/Shire, BMS, Celgene, Chugai, Janssen-Cilag, MSD, Novartis, Pfizer, Roche, UCB, Ina Koetter: None declared, Claudia Guenther Grant/research support from: Pfizer, Novartis, Employee of: 20 years ago, Novartis, Speakers bureau: Celtis, Tim Schmeiser Speakers bureau: Actelion, UCB, Pfizer, Gabriele Zeidler Grant/research support from:

- MSD Sharp & Dohme GmbH
- Actelion Pharmaceuticals Deutschland GmbH
- Roche Pharma AG
- AbbVie Deutschland GmbH Co. KG
- Pfizer Pharma GmbH
- Lilly Deutschland GmbH
- Celgene GmbH
- Bristol-Myers Squibb

UCB Pharma GmbH/UCB GmbH

- Speakers bureau:
- MSD Sharp & Dohme GmbH
- Actelion Pharmaceuticals Deutschland GmbH
- Roche Pharma AG
- AbbVie Deutschland GmbH Co. KG
- Pfizer Pharma GmbH
- Lilly Deutschland GmbH
- Celgene GmbH
- Bristol-Myers Squibb
- UCB Pharma GmbH/UCB GmbH

Disclosure of Interests: Alexander Kreuter Speakers bureau: Actelion Pharma, MSD, AbbVie, InfektioPharm, Margitta Worm: None declared, Laura Susok: None declared, Aaron Juche: None declared, Christiane Pfeiffer: None declared, Cord Sunderkoetter: None declared, Donja Homayoon: None declared, Nicolas Hunzelmann Speakers bureau: Boehringer Ingelheim, Actelion, Pfizer, Roche

DOI: 10.1136/annrheumdis-2019-eular.1788

AB0216 THE IMBALANCE OF ACTIVITIES OF PURINE AND PYRIMIDINE METABOLISM ENZYMES IN RED BLOOD CELLS OF SYSTEMIC SCLERODERMA PATIENTS

Elena Mozgovaya, Svetlana Bedina, Andrew Trofimenko, Irina Zborovskaya, Maria Mamus, Ekaterina Tikhomirova. *Federal State Budgetary Institution "Research Institute of Clinical and Experimental Rheumatology named after A.B. Zborovskiy", Volgograd, Russian Federation*

Background: The systemic sclerosis (SSc) is connective tissue disease of unknown cause. The key points in development of SSc are the increased number of high collagen-producing fibroblasts in the skin, endothelial dysfunction and immune activation. Recent studies report various metabolic disturbances in SSc patients. Purine and pyrimidine metabolic pathways are underlies the central processes of cellular life. The changes in activity of purine and pyrimidine metabolism enzymes in blood plasma and lysed lymphocytes depending on the SSc activity were described by us earlier [1]. At the same time, there are publications that confirm the relationship between changes in ADA activity in red blood cells and pronounced immune disorders [2].

Objectives: to characterize enzymatic patterns of the major purine and pyrimidine metabolic pathways enzymes in lysed red blood cells depending on the SSc activity.

Methods: 51 SSc patients and 30 healthy controls were enrolled in the study. The diagnosis was verified in accordance with the international standards (ACR/EULAR 2013). Disease activity was assessed in accordance with the national classification [3]. Adenosine deaminase (ADA; EC 3.5.4.4); adenosine kinase (AK; EC 2.7.1.20); guanylate kinase (GK; EC 2.7.4.8), dihydroorotate dehydrogenase (DODH; DC 1.3.1.14); IMP

dehydrogenase (IMPDH; EC 1.1.1.205); purine nucleoside phosphorylase (PNP; EC 2.4.2.1); thymidine kinase (TK; EC 2.7.1.21); thymidine phosphorylase (TP; EC 2.4.2.4); uracil/thymidine dehydrogenase (UDH; EC 1.17.99.4); cytidine deaminase (CDA; EC 3.5.4.5) activities were measured in lysed red blood cells.

Results: Mean age of patients (Mean± SD) was 42.8±1.3years, mean SSc duration was 7.9±0.7 years. We revealed substantial changes in enzymatic activities related to both purine and pyrimidine metabolism in lysed red blood cells of SSc patients. The increased ADA (p<0.001), AK (p<0.001), IMPDH (p<0.001), TK (p<0.001), UDH (p<0.001) activities and the decreased DODH (p<0.001), PNP (p<0.001) activities in lysed red blood cells were observed of SSc patients in comparison with healthy controls. AK, IMPDH, TK, UDH activities positively correlated with SSc activity. Negative correlations with SSc activity were revealed for ADA, CDA, DODG, GK, PNP, TP activities.

Conclusion: The progression of SSc goes with the imbalance of the purine and pyrimidine enzymes in a regular manner. Activity of the autoimmune inflammation is the factor that underlies the enzymatic pattern of purine and pyrimidine metabolism.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2019-eular.2125

AB0217 ASSOCIATION OF EUROPEAN SCLERODERMA STUDY GROUP ACTIVITY INDEX (EScSG-AI) WITH PROGRESSION PULMONARY ALTERATIONS BY HRCT IN PATIENTSWITH SYSTEMIC SCLEROSIS OVER A FIVE YEAR PERIOD

Olga Ovsyannikova, Lidia P. Ananyeva, Olga Koneva, Liudmila Garzanova, Oxana Desinova, Mayya Starovoytova. *V.A. Nasonova Research Institute of Rheumatology, laboratory of microcirculation and inflammation, Moscow, Russian Federation*

Background: Systemic sclerosis (SSc) is a rare connective tissue disease with a heterogeneous clinical course. Interstitial lung disease (ILD) is a common manifestation of SSc and a leading cause of death. Patients with early active SSc are at great risk for progressive ILD.

Objectives: To assess the EScSG-AI in patients with systemic sclerosis (SSc) and interstitial lung disease over a five year period.

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. Disease activity was assessed by the 2001 European Scleroderma Study Group Activity Index (EScSG- AI).

Results: There were no significant differences between groups related to sex, frequency of diffuse form and duration disease. Mean dates of EScSG-AI score of all pts and in the each groups in first visit and the end of follow up are present in table 1.

Table 1

Groups	First visit	At the end of the study	p
Group 1 (n=16)	1.9 ± 1.8	1.7 ± 0.9	P > 0.05
Group 2 (n=39)	2.0 ± 1.5	2.1 ± 1.3	P > 0.05
Group 3 (n=22)	2.4 ± 1.5	3.25 ± 2.0	P < 0.05
All pts (n= 77)	2.2 ± 1.6	1.9 ± 1.8	P > 0.05

After 5 years of follow up mean values of EScSG-AI score increased significantly in group 3 and was more than 3, this actually means the activity of the disease. The mean values of EScSG-AI score in group 3