References:
[1] Selva-O'Callaghan, A., Pinal-Fernandez, I., Trallero-Araguás, E., Milisenda, J. C., Grau-Junyent, J. M., & Mammen, A. L. (2018). *Classification and management of adult inflammatory myopathies. The Lancet Neurology, 17(9), 816–828.*
[2] Wu C, Wang Q, He L, Yang E, Zeng X (2018) *Hospitalization mortality and associated risk factors in patients with polymyositis and dermatomyositis: A retrospective case-control study. PLoS ONE 13(2): e0192491.*
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.3694

AB0566 NAILFOLD EXTENT OF REDUCED CAPILLARY DENSITY IS ASSOCIATED WITH DIGITAL ULCERS AND WITH AN INCREASED RISK OF DIGITAL ULCERS IN SYSTEMIC SCLEROSIS

R. De Angelis[1], F. Salaffi[1]. [1]*Marche Polytechnic University, Department of Clinical and Molecular Sciences, Ancona, Italy*

Background: A growing evidence supports the role of microvasculopathy as a primary pathogenic event in systemic sclerosis (SSc). The most commonly used imaging technique to identify microangiopathy in SSc is high magnification videocapillaroscopy (NVC), and reduced capillary density and/or capillary loss, which is a typical feature of "scleroderma microangiopathy", easily identified by NVC, has been associated with digital ulcers (DUs). Different approaches have been proposed to measure capillary density or capillary loss. Some of these were qualitative methods, others semi-quantitative, others only concerned a limited nailfold area, without ever evaluating the overall density, which is more suitable for quantitative estimate.
Objectives: To assess the association between the extent of different values of nailfold capillary density and the presence of DUs and to identify the risk of developing DUs, based on quantitative parameters.
Methods: The study involved 54 SSc selected patients (47 women and 7 men, mean age 59.5 years, 50 with limited and 4 with diffuse). The study population came from an ongoing database, that includes clinical and laboratory data of patients with definite SSc. A videocapillaroscope (VideoCap® 3.0, DS Medica, Milan, Italy) with a 200x optical probe was used. During examination, eight fingers (fingers 2–5 of each hand), 4 fields per finger, according to the standard literature were assessed. For each patient, a total of 32 images were collected, then classified as having either "normal", "non-specific" or the "scleroderma pattern" (SP). Capillary density was defined as the number of capillaries/mm in the distal row, regardless of its shape and morphology. Avascular areas were defined by the absence of loops within a width/area extending over more than 500 microns. For each patient, the SP images were further graded with no/slight reduction of the capillary density (7-9 loops/mm) (NOR), with a well-defined reduction of capillary density (6-4 loops/mm) (RED) and with loss of capillaries (<4) plus avascular areas (AA). Then, the overall percentages were calculated (the number with SP, the number with NOR, with RED and with AA, respect to 32), thus obtaining the quantitative measures. All data were analysed using the MedCalc® version 18.6; 64-bit (MedCalc Software, Mariakerke, Belgium).
Results: A total of 1728 images were analyzed. Patients with DUs were 16/54 (29.6%). All patients had a SP, but only five patients showed a SP along the entire nailfold. A comparison between patients with or without DUs showed a significant difference both for the overall extent of AA (p=0.032), and particularly for the overall extent of RED (p<0.001). No significant difference was found regarding the overall extent of the SP (p=0.085). Factor significantly associated with DUs in multivariate analysis was the overall extent of RED (p=0.0286). The ROC curve was very effective at discriminating the capillary feature able to distinguish patients with DUs from patients without DUs. The discriminatory power of the overall extent of RED was very good, with an AUC of 0.948 (95 % CI 0.852 ± 0.990). Then, we calculated the cut-off values of the overall extent of RED for presence/absence of DUs with the highest combination of sensitivity and specificity. The resulting cut-off value (Yourden index of 0.825) was >68.7 (sensitivity 92.31 %; specificity 90.24 %) with a LR+ of 9.46.
Conclusion: Our data strongly support that the capillary density between 4 and 6 loops/mm is the best capillaroscopic quantitative measure associated with DUs and able to discriminate the probability of having DUs. If all SSc-specific antibodies and/or other laboratory/clinical parameters are not yet available, the overall capillary density can allow physicians to assess SSc patients easily, regarding DUs and risk for developing DUs.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.4173

AB0567 CHARACTERIZATION OF PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHY AND MYOCARDIAL INVOLVEMENT: A MONO-CENTRIC EXPERIENCE.

R. De Lorenzo[1,2], S. Cavalli[2], F. Bonomi[2], S. Tronci[1,2], S. L. Calvisi[1], S. Previtali[1], P. Rovere-Querini[1,2]. [1]*San Raffaele Hospital, Milan, Italy*; [2]*San Raffaele University, Milan, Italy*

Background: Idiopathic inflammatory myopathies (IIM) are immune-mediated disorders of the skeletal muscle, with dermatomyositis (DM), polymyositis (PM), inclusion body myositis (IBM) and immune-mediated necrotizing myopathy (IMNM) representing major subtypes. Beyond skeletal muscle, other organs may be affected and myocardial involvement may lead to severe life-threatening complications. The exact prevalence of myocardial involvement among IIM patients and its impact on other disease characteristics remain unclear.
Objectives: To investigate the prevalence of myocarditis in patients affected by IIM in and to determine whether the presence and extent of myocardial involvement identify a distinct disease phenotype.
Methods: 42 longitudinally followed IIM patients were routinely screened for myocardial involvement during a median [IQR] follow-up time of 4.2 [2-8.5] years. Patients with secondary causes of myocardial dysfunction were not included. Patients were considered to have myocarditis in case of: *i)* abnormal elevation of both circulating troponin T and troponin I, *ii)* signs of myocardial inflammation or necrosis/fibrosis at cardiac MRI, or *iii)* positive myocardial tissue histology. Demographic, clinical and serologic features of patients with myocarditis were compared to those with no sign of myocardial involvement. Moreover, we determined whether the extent of myocardial involvement based on troponin levels predicts skeletal muscle disease severity.
Results: 57.1% (24 of 42) of patients had myocarditis. The frequency of myocardial dysfunction was similar among patients with DM, PM, IBM or IMNM and was not related to autoantibody positivity. Myocarditis was not associated with sex or ethnicity. Patients with or without myocarditis were similar in terms of age at disease onset and extra-muscular manifestations including dysphonia, dysphagia, arthralgias or arthritis, Raynaud phenomenon or interstitial lung disease. Independent of the IIM subtype, the presence of perimysial macrophages at skeletal muscle biopsy seems to protect from myocarditis development (p=0.04). Patients with myocarditis had higher median [IQR] levels of aldolase (10.9 [7.8-15.8] vs. 5.6 [4.9-8.6], p=0.014) and creatine kinase (1785 [966-5852] vs. 685 [168-2255], p=0.04) compared to patients with no myocardial dysfunction. Among patients with myocarditis, levels of troponin I negatively correlated with manual muscle testing 8 (MMT8) score (r=-1, p=0.01), strength in biceps (r=-0.95, p=0.014) and wrist extensors (r=-0.95, p=0.014) at last visit. Troponin T and troponin I titers were similar among patients with different IIM subtypes. C-reactive protein (p<0.04) but not erythrocyte sedimentation rate was found to predict myocardial involvement.
Conclusion: Our findings suggest that myocarditis is a frequent occurrence among patients with IIM and should be routinely ruled out. A more severe skeletal muscle disease is associated with an increased likelihood of myocarditis development, presumably due to higher systemic disease activity or inefficient disease control. The extent of myocardial damage faithfully reflects the severity of skeletal muscle dysfunction.
References:
[1] Dalakas MC. Inflammatory muscle diseases. N Engl J Med. 2015 Apr 30;372(18):1734-47. doi: 10.1056/NEJMra1402225.
[2] Schwartz T, Diederichsen LP, Lundberg IE, et al. Cardiac involvement in adult and juvenile idiopathic inflammatory myopathies. RMD Open 2016;2:e000291. doi:10.1136/rmdopen-2016- 000291.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.6248

AB0568 BASELINE EUROPEAN PATIENT DEMOGRAPHICS AND DISEASE CHARACTERISTICS IN A PHASE 3 STUDY OF SAFETY AND EFFICACY OF LENABASUM, A CB2 AGONIST, IN DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

C. Denton[1], B. Bloom[2], N. Dgetluck[2], B. White[2], R. Spiera[3]on behalf of RESOLVE-1 Study Investigators. [1]*University College London, London, United Kingdom*; [2]*Corbus Pharmaceuticals, Inc., Norwood, United States of America*; [3]*Hospital for Special Surgery Main Hospital, New York, United States of America*

Background: We previously presented on the baseline characteristics of a large cohort of diffuse cutaneous systemic sclerosis (dcSSc) patients enrolled in a Phase 3 trial of lenabasum, a selective cannabinoid receptor type 2 (CB2) agonist. Lenabasum, was safe and well-tolerated in a prior Phase 2