had an average of 0.95 (mild disability) versus 0.77 in the RA group, and the average hand function score or Cochin index: 20.6 (moderate disability) against 17.5 of the RA group. There was no statistically significant correlation (χ^2) between the elevated CRP values and these 2 questionnaires in the SS group. The radiological findings of calcinosis and acroosteolysis in the group with SS were double the cases of the group with RA (7 versus 3, and 14 versus 7 respectively).

Conclusions: Subclinical joint involvement in systemic sclerosis had a high prevalence, similar in rheumatoid arthritis, with ultrasound being a fundamental test when evaluating subclinical inflammatory activity. It also presents an important correlation with the findings found in radiographs in the same locations.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7102

FRI0467

THE RELATIONSHIP BETWEEN ^{99m}Tc-PERTHNETATE HAND PERFUSION SCINTIGRAPHY AND NAILFOLD CAPILLOSCOPY IN SYSTEMIC SCLEROSIS PATIENTS: A PILOT STUDY

S.R. Pavlov-Dolijanovic¹, N. Petrovic², N. Vujasinovic Stupar¹, N. Damjanovic¹, G. Radunovic¹, V. Zugic³, D. Babic⁴, D. Sobic-Saranovic², V. Artiko². ¹School of Medicine, University of Belgrade, Institute of Rheumatology, Belgrade, Serbia; ²School of Medicine University of Belgrade, Center for Nuclear Medicine, Clinical Center of Serbia, Belgrade, Serbia; ³School of Medicine, University of Belgrade, Clinic for Pulmology, Clinical Center of Serbia, Belgrade; ⁴School of Medicine University of Belgrade, Institute for Medical Statistics and Informatics, Belgrade, Serbia, Belgrade, Serbia

Background: The aim of this study was to assess possible relationship between the altered blood perfusion at hands analysed by ^{99m}Tc-pertechnetate hand perfusion scintigraphy (^{99m}TcPHPS) and, morphological microvascular abnormalities detected by nailfold capillaroscopy (NC) in SSc patients.

Methods: The study group consisted of 25 patients with SSc (14 with diffuse SSc, and 11 with limited SSc) and 28 control subjects (18 patients with primary RP, and 10 healthy individuals). NC and ^{99m}TcPHPS was performed in all the groups examined. The capillaroscopic pattern was classified as normal or scleroderma ("early", "active", or "late") pattern. Gamma-camera dynamic first-pass study during the first 60 s and a static scintigraphy after 5 min were recorded following a bolus injection of ^{99m}Tc-pertechnetate via a cubital vein. Regions of interest were drawn on the summed images around the fingers and the palmar region. The fingers-to-palm ratios were then calculated.

Results: SSc patients showed a significantly lower blood flow (BF) and blood pool (BP), (0.43 ± 0.21 vs 0.36 ± 0.07 , respectively), than PRP patients (0.45 ± 0.18 vs 0.42 ± 0.06 , respectively) and healthy subjects (0.58 ± 0.19 vs 0.44 ± 0.06 , respectively), (p-value 0.039 vs 0.004, respectively). A gradual decrease of BF and BP was found in SSc patients with progressive severity of NVC patterns of microangiopathy ["early" (0.49 ± 0.03 vs 0.39 ± 0.04 , respectively), "active" (0.43 ± 0.11 vs 0.38 ± 0.06 , respectively) or "late" (0.40 ± 0.28 vs 0.36 ± 0.08 , respectively), (p-value 0.462 vs 0.728 respectively), but these differences were not statistically significant. Patients with diffuse SSc showed lower BF, and higher BP (0.42 ± 0.26 vs 0.37 ± 0.07 , respectively) than those with limited SSc, (0.44 ± 0.14 vs 0.35 ± 0.064 , respectively), but this differences is not statistical significantly (p=0.76 vs p=0.53, respectively). There was no significant correlation between BF and BP values and type of SSc (limited or diffuse) ($r=-0.06$, p=0.77; $r=0.13$, p=0.54, respectively) as well as three microangiopathy patterns ($r=-0.253$, p=0.22; $r=-0.13$, p=0.54, respectively).

Conclusions: NC represents the best method to analyse microvascular damage in rheumatic diseases, especially SSc. ^{99m}TcPHPS improves the evaluation of vascular damage in SSc patients. There is no direct relationship between these two methods, but one method complements another in the study of vascular damage in SSc patients.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4268

FRI0468

ABNORMAL OESOPHAGEAL MOTILITY DURING A SOLID TEST MEAL IN SYSTEMIC SCLEROSIS – DETECTION IN VERY EARLY DISEASE AND ASSOCIATION WITH DISEASE PROGRESSION

S. Bütikofer¹, S. Jordan², M. Sauter¹, M. Hollenstein¹, H. Heinrich¹, N. Freitas-Queiroz³, T. Kuntzen¹, P. Valli¹, D. Ang⁴, M. Oberacher⁵, B. Maurer², W. Schwitler¹, M. Fox¹, O. Distler², B. Misselwitz¹. ¹Department of Gastroenterology, ²Department of Rheumatology, University Hospital Zurich, Zurich, Switzerland; ³Department of Gastroenterology, University of Sao Paulo, School of Medicine, Sao Paulo, Brazil; ⁴Department of Gastroenterology, Changi General Hospital, Singapore, Singapore; ⁵Department of Medicine, Spital Limmattal, Schlieren, Switzerland

Background: Ineffective oesophageal motility (IEM) is frequent in patients with systemic sclerosis (SSc). High-resolution oesophageal manometry (HRM) is the reference standard test for oesophageal motility and addition of a test meal increases diagnostic sensitivity and specificity.

Objectives: This study assessed whether using a test meal instead of standard water swallow in HRM increases sensitivity and can detect clinically relevant, abnormal motility in already very early SSc and whether this finding is associated with subsequent disease progression.

Methods: This prospective, longitudinal cohort study recruited 68 consecutive SSc patients (group #1: 32 established disease (ACR/EULAR 2013 and ACR 1980 criteria fulfilled); group #2: 24 early disease (only ACR/EULAR 2013 fulfilled); group #3: 12 very early disease (clinical expert diagnosis of SSc, no classification criteria fulfilled) and 72 healthy controls. HRM evaluated oesophageal motility for water swallows and a solid test meal using validated methods.

Results: SSc patients had less frequent effective oesophageal contractions during the test meal compared to healthy controls. Notably, this was detected even in very early disease (0.15, 1.0, 2.1/min for group #1, #2 and #3, vs. 2.5/min in health, p<0.001; p<0.001 and p<0.009, respectively). No other significant abnormality on HRM was found in patients with very early disease (group 1). Ineffective motility at HRM was associated with a higher modified Rodnan skin score at baseline. Moreover, at mean 18¹⁰⁻³¹ months follow-up, the presence of ineffective motility at baseline was associated with progression of skin disease for the overall SSc cohort (p<0.010). In a secondary analysis, below-average lower oesophageal sphincter pressure was associated with progression of skin disease and organ disease, in particular interstitial lung disease (p<0.009).

Key HRM findings in SSc patients and healthy volunteers.

A: Percentage of effective swallows in the test meal. **B:** Number effective swallows per min in the test meal. **C:** Number pharyngeal swallows per min in the test meal. **D:** Vigour of peristalsis (DCI) in standard HRM. Median (red bar) and IQR (error bars) are indicated. Please, note the significant decrease in effective swallows per minute which in patients with very-early disease, which is only observed for the test meal. Statistical analysis: Mann-Whitney U test. * p<0.05, ** p<0.01, *** p<0.001.

Conclusions: Ineffective motility during a test meal is present already in patients with very early SSc. In cross-sectional analysis, findings on HRM studies at baseline are associated with disease severity and prospectively with progression of

skin disease during follow-up. Thus, performance of HRM already in very early disease stages can support individual risk-stratification of SSc patients.

Disclosure of Interest: S. Bütikofer Consultant for: Consultancies<\$10000, Speakers bureau: Speaking fees<\$10000, S. Jordan: None declared, M. Sauter: None declared, M. Hollenstein: None declared, H. Heinrich: None declared, N. Freitas-Queiroz: None declared, T. Kuntzen Consultant for: Consultancies<\$10000, Speakers bureau: Speaking fees<\$10000, P. Valli: None declared, D. Ang: None declared, M. Oberacher: None declared, B. Maurer: None declared, W. Schwitzer: None declared, M. Fox Grant/research support from: Nestle Research International, AstraZeneca R and D, Given Imaging, and Reckitt Benckiser, Consultant for: Given Imaging, AstraZeneca, Reckitt Benckiser, Shire, Almirall and Sucampo., Speakers bureau: Given Imaging, Medical Measurement Systems and Sandhill Scientific Instruments., O. Distler Grant/research support from: Actelion, Bayer, Boehringer Ingelheim, Mitsubishi Tanabe Pharma and Roche, Consultant for: Actelion, Bayer, BiogenIdec, Boehringer Ingelheim, ChemomAb, espeRare foundation, Genentech/Roche, GSK, Inventiva, Italfarmaco, Lilly, medac, MedImmune, Mitsubishi Tanabe Pharma, Pharmacyclis, Novartis, Pfizer, Sanofi, Sinova and UCB, B. Misselwitz Speakers bureau: Speaking fees<\$10 000 for Given Imaging

DOI: 10.1136/annrheumdis-2018-eular.3030

FRI0469

ACUTE AND CHRONIC EFFECTS OF TWO DIFFERENT INTRAVENOUS ILOPROST REGIMENS IN SYSTEMIC SCLEROSIS: A SINGLE CENTRE PRAGMATIC NON-RANDOMISED TRIAL

T. Schioppo¹, A. Orenti^{2,3}, P. Boracchi^{2,3}, O. De Lucia¹, A. Murgio¹, F. Ingegnoli^{1,2}

¹Division of Rheumatology, ASST Pini-CTO; ²Dept of Clinical Sciences and Community Health, Università degli Studi di Milano; ³Lab of Medical Statistics, Epidemiology and Biometry GA Maccacaro, Milano, Italy

Background: In systemic sclerosis (SSc) vascular involvement plays a pivotal role in the pathogenesis and clinical manifestations.¹ Intravenous (IV) iloprost (ILO), a prostacyclin analogue, is administered in SSc, according to EULAR recommendations, to improve Raynaud's phenomenon (RP) and to achieve digital ulcers (DU) healing and prevention,² even though no further indications about the regimen are specified.

Objectives: The aim of this study was to evaluate acute and chronic effects of IV ILO administered with two different regimens as assessed by power Doppler ultrasound (PDUS) and nailfold videocapillaroscopy (NVC) and in a group of patients not treated with ILO.

Methods: This was a single centre pragmatic non-randomised trial on 96 patients with SSc divided in 3 groups: no ILO (group A), ILO once monthly (group B) and ILO for 5 consecutive days every 3 month (group C). At each evaluation patients underwent PDUS and NVC. Group A patients were examined at baseline and after 3 months. Group B every month before and after IV ILO monthly therapy for 3 months. Group C before and after the 1 and 5 days of therapy at baseline and after 3 months. PDUS parameters included resistivity index (RI), finger pulp blood flow (FPBF) and periungual vascularisation. The sum of capillaries apex width in one millimetre was assessed through NVC. Results were analysed considering the average outdoor temperature at the place of residence.

Results: An acute IV ILO effect was observed for FPBF in group B and C ($p < 0.001$ and 0.005 respectively). An acute effect was observed for RI and periungual vascularisation only in group B. A progressive increase was observed for the other parameters without being statistically different. On the contrary IV ILO effects were not observed any longer before the following infusion. Moreover, some parameters (FPBF in group B and RI in group C) showed a statistically higher increase as low as the outdoor temperature was.

Conclusions: IV ILO therapy was able to cause an acute effect with respect to PDUS parameters, especially in group B. The acute effect was not any longer maintained until the following infusion. Future studies are needed to assess time for re-treating.

REFERENCES:

- [1] Matucci-Cerinic M, et al. Review: evidence that systemic sclerosis is a vascular disease. *Arthritis Rheum.* 2013;65(8):1953–62.
- [2] Kowal-Bielecka, et al. Update of EULAR recommendations for the treatment of systemic sclerosis. *Ann Rheum Dis.* 2017;76(8):1327–1339.

Acknowledgements: We thank Meteo Operations Italia (MOPI) Srl – Centro Epson Meteo for providing temperature data.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.1783

FRI0470

A PHASE 2 STUDY OF SAFETY AND EFFICACY OF LENABASUM (JBT-101), A CANNABINOID RECEPTOR TYPE 2 AGONIST, IN REFRACTORY SKIN-PREDOMINANT DERMATOMYOSITIS

V.P. Werth^{1,2}, E. Hejazi^{1,2}, S.M. Pena^{1,2}, J.S. Haber^{1,2}, J. Okawa^{1,2}, R. Feng^{1,2}, K. Gabre², J.S. Concha^{1,2}, C. Cornwall³, N. Dgetluck³, S. Constantine³, B. White³.
¹Philadelphia Veteran Affairs Medical Center, ²University of Pennsylvania, Philadelphia; ³Corbus Pharmaceuticals, Inc., Norwood, USA

Background: Effective treatment options are limited for refractory skin disease in dermatomyositis (DM). Lenabasum is a non-immunosuppressive, synthetic, oral preferential CB2 agonist that triggers resolution of innate immune responses and reduces cytokine production by PBMC from DM patients.

Objectives: The purpose of this study was to test safety and efficacy of lenabasum (aka JBT-101, anabasum) in DM subjects with refractory, moderate-to-severely active skin disease.

Methods: A double-blind, randomised placebo-controlled 16 week Phase 2 trial (JBT101-DM-001; NCT02466243) enrolled adults with documented DM and a Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) activity score ≥ 14 , minimal active muscle involvement and failure or intolerance to hydroxychloroquine and stable DM medications including immunosuppressants. Subjects received 2 escalating dose levels of lenabasum (20 mg QD X 4 weeks, then 20 mg BID X 8 weeks) or PBO X 12 weeks. Subjects were followed off study drug X 4 weeks. Safety and efficacy outcomes were assessed from Day 1 through end of study at Week 16. The primary efficacy objective was to assess efficacy of lenabasum using CDASI activity score.

Results: 11 adults each received lenabasum and PBO (n=22). Demographic and disease characteristics were similar in both cohorts. Both cohorts had mean CDASI activity scores in the severe range^{33–35} despite immunosuppressants (19/22 subjects). Lenabasum subjects had clinically meaningful improvement in CDASI activity scores with mean reduction ≥ 5 points at all visits after 4 weeks. Improvement had statistical significance at end of study (figure 1, $p=0.02$, 2-sided MMRM) that first became apparent after 4 weeks. Lenabasum provided greater improvement than placebo in CDASI damage index, patient-reported global skin disease and overall disease assessments, skin symptoms including photosensitivity and itch, fatigue, sleep, pain interference with activities, pain, and physical function (examples in figure 1). Improvements in secondary efficacy outcomes reached statistical significance ($p \leq 0.1$, 1-sided MMRM) at multiple visits after week 4 (figure 1). There were no serious, severe or unexpected adverse events (AEs) related to lenabasum. Tolerability of lenabasum was excellent with no study drop-outs. Subjects in the lenabasum cohort had numerically more mild AEs than placebo subjects (29 vs. 19) and fewer moderate AEs (4 vs. 7). AEs in ≥ 3 subjects in any cohort were diarrhoea, dizziness (lightheadedness), fatigue and dry mouth.

Abstract FRI0470 – Figure 1. Effects of Lenabasum on Efficacy Outcomes in Refractory, Skin-Predominant DM

Conclusions: Lenabasum demonstrated consistent evidence of clinical benefit across multiple efficacy outcomes and had acceptable safety and tolerability in this Phase 2 trial in refractory skin disease in DM. Further evaluation of lenabasum in the treatment of DM is warranted.

Disclosure of Interest: V. Werth Consultant for: Corbus Pharmaceuticals, Inc., E. Hejazi: None declared, S. Pena: None declared, J. Haber: None declared, J. Okawa: None declared, R. Feng: None declared, K. Gabre: None declared, J. Concha: None declared, C. Cornwall Employee of: Corbus Pharmaceuticals, Inc., N. Dgetluck Employee of: Corbus Pharmaceuticals, Inc., S. Constantine Employee of: Corbus Pharmaceuticals, Inc., B. White Employee of: Corbus Pharmaceuticals, Inc.

DOI: 10.1136/annrheumdis-2018-eular.3531