chest CT (10/18) before and after treatment. In particular, we considered PFTs performed 6 months before RTX, at time 0, one year after the first RTX cycle, and at the end of follow-up (13.7±7.3 years). ILD extent score was assessed by the semi-quantitative method proposed by Goh et al (2008).[1]

Results: Forced vital capacity (FVC%) significantly reduced during the year before RTX treatment [from 95.2±17.4 to 84.8±16.4; p=0.0017], as well as the diffusing capacity for carbon monoxide (DLCO%) [from 58.1±14.3 to 47.6±12.9; p=0.0002]. Conversely, FVC% and DLCO% stabilised one year after the first RTX cycle (80.8±23 and 47.8±15.7, respectively), and at the end of the follow-up (84.3 ±24.6 and 54.8.8±12.3, respectively; p=0.0001). In our cohort, only 7/10 patients had ILD detectable on CT before treatment. At the end of the follow-up we observed that ILD extented in 6/7 cases one patient remained stable, the three subjects without ILD did not developed pulmonary fibrosis.

Conclusions: According to PFTs results, our study showed that RTX could stabilise the progression of lung function tests in SSc patients. However, the semi-quantitative visual score identified radiological pulmonary worsening in many patient with stabilised PFTs. Therefore, the correlations between functional and radiological outcomes are so weak that many Authors suggested they should be considered together in SSc-ILD assessment.

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FRI0443 DIGITAL ARTERY VOLUME INDEX: THE FIRST OBJECTIVE, AUTOMATED, NON-INVASIVE IMAGING DIAGNOSTIC OF MACROVASCULAR INVOLVEMENT IN SSC

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Background: Macrovascular involvement in Systemic Sclerosis (SSc) is caused by proliferation of vascular smooth muscle cells within the intima of arteries (neointimal proliferation). The resulting decrease in arterial volume is responsible for the severity of Raynaud's and for the onset of severe clinical manifestations such as Renal Crisis and Pulmonary Artery Hypertension.

Several RCTs have demonstrated the efficacy of Endothelin Receptor Antagonists (ERAs) in targeting neointimal proliferation, which makes them to date, the only disease modifying agent available in SSc. Nevertheless, the lack of validated early diagnostics means the intention to treat with ERAs remains limited to the diagnosis of end stage clinical manifestations of neointimal proliferation, such as presence of Digital Ulcer (DU) and PAH.

Objectives: Here we aimed to determine the proof of concept validity of automated Digital Artery Volume Index (DAVIX) measured by non-contrast Time of Flight (TOF) MRI as an objective diagnostic tool to be used as surrogate outcome measure of neointimal proliferation in SSc.

Methods: 10 Healthy Volunteers (HV) and 8 SSc patients were enrolled. Six patients underwent longitudinal assessments at least 12 months apart. MRI scans were performed on a 3T Magnetom Verio (Siemens) and consisted of a VIBE 3D T1 scan and a 2D TOF sequence of 8 min. DAVIX was calculated as the percentage of the ratio of digital artery and the respective finger volumes. The vessels and fingers were manually outlined by an expert radiologist and used as a 'gold standard' (DAVIX-1) and compared to a 2nd independent radiologist assessment (DAVIX-2). An automated segmentation algorithm was developed using threshold based segmentation and region growing (DAVIX-A) and validated against gold standard. Intraclass correlation coefficient (ICC) and absolute agreement were calculated to estimate reliability. Bland-Altman bias and 95% limits of agreement (LoA) were calculated as well.

Results: SSc patients and HV had comparable age (44±8 vs 45±8) and gender (F:M=5:2 vs 4:2). 4 fingers were affected by DUs at baseline and two fingers were affected by new DU at follow up. Mean(±SD) DAVIX in HV was 1.21 (±0.39) with no significant difference among individual fingers. Mean DAVIX in SSc patients was 0.37 (±0.18) and 0.32 (±0.39) at baseline and follow-up, respectively (p<0.0001 vs HV for both). The fingers with DU had an average DAVIX of 0.24 (±0.08) vs 0.40 (±0.18) of the fingers without DU (p=0.02). DAVIX changed over time in both directions. In the fingers affected by new DU, DAVIX dropped by 56% (0.161 vs 0.072) and 86% (1.149 vs 0.132), respectively. ICC among two independent readings was 0.90 (95% LoA −0.139, 0.118) with overall r^2=0.74 (p<0.0001). Automated segmentation showed superior correlation to gold standard with r^2=0.80 (p<0.0001).

Conclusions: This proof of concept study demonstrated validity and sensitivity to change of DAVIX for the automated volumetric assessment of digital arteries, which reflected clinical worsening in patients with new DU. Larger, longitudinal studies are planned to assess DAVIX' predictive value for the onset of DU and its potential use as early diagnostic of neointimal proliferation in SSc.

Disclosure of Interest: None declared

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FRI0444 PERIOSTIN IN SYSTEMIC SCLEROSIS: SERUM LEVELS AND SKIN EXPRESSION OF A NOVEL POSSIBLE BIOMARKER

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Background: Periostin(PN), a matricellular protein, serves as a regulator of wound healing and fibrosis. $PN^{-/-}$ mice develop lower degrees of fibrosis when exposed to bleomycin(BLM), suggesting a possible role in the pathogenesis of Systemic Sclerosis(SSc)[1]. PN serum levels are increased in SSc and seem to be associated with skin disease severity[2].

Objectives: To evaluate the role of serum PN as a biomarker of SSc severity, and to determine PN tissue expression in SSc patients.

Methods: PN serum levels were assessed by ELISA in 48 patients: 3 primary Raynaud's, 12 early SSc, 11 SSc without organ involvement, 22 SSc with organ involvement. All SSc patients met 2013 ACR/EULAR criteria. PN serum levels were evaluated in 28 sex-/age-matched healthy-controls(HCs). Data regarding disease subtypes and organ involvement were correlated. PN skin expression was determined by immunohistochemistry on paired involved and uninvolved skin biopsy samples in 10 patients(4 lcSSc and 6 dcSSc) in combination with a-SMA, CD68, CD3, CD4, CD8, CD163, CD20 and CD131.

Results: PN serum levels were higher in SSc patients compared to HCs(32.7 ±8.0 vs 27.7±7.3 ng/ml, p<0.001). Its levels were comparable among different groups. No differences in PN serum levels were detected when comparing disease subtypes, disease duration, presence and extent of organ involvement, autoantibodies profile and current or previous treatment. Higher PN levels were found in SSc patients with active pattern at nailfold videocapillaroscopy and a history of digital ulcers(p<0.02). PN serum levels did not correlate with skin or lung disease extent. Skin samples from involved SSc skin showed high PN expression in the upper dermis and in the fibrotic area of the lower dermis(more evident in dcSSc), suggesting a role in skin fibrosis. In all SSc involved skin, PN was expressed in areas where ongoing fibroproliferation and macrophage/T lymphocytic infiltration occured, indirectly suggesting a pathogenic role in inflammation driven-fibrosis. Interestingly, an identical PN immunohistochemical expression was evident in uninvolved dcSSc skin, but not in lcSSc skin.

Conclusions: In our cohort PN serum levels are elevated in SSc patients but they do not correlate with disease features. Its postulated role as a severity biomarker needs to be further elucidated. The different immunohistochemical expression of PN in uninvolved skin from dcSSc and lcSSc patients suggests a possible pathogenic role in the progressive inflammation-driven fibrosis that characterise diffuse cutaneous involvement.

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FRI0445 A RANDOMISED CONTROLLED TRIAL TO COMPARE THE EFFICACY OF ORAL MYCOPHENOLATE MOFETIL WITH PLACEBO IN PATIENTS WITH SYSTEMIC SCLEROSIS RELATED EARLY INTERSTITIAL LUNG DISEASE

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Background: Previous studies showed benefit of immunosuppressants in moderate to severe ILD in systemic sclerosis (SSc).[1,2] Initiation of immunosuppression early in the course of SSc-ILD might help in halting disease process and improve long term morbidity and mortality.

Objectives: Aim of the study was to determine efficacy and safety of mycophenolate mofetil (MMF) in treating early and mild SSc-ILD (ILD on HRCT, FVC ≥70%