sclerosis (SSc) and it substantially contributes to prominent features of the disease such as digital ulcers (DUs). History of DUs (HDUs) has been shown to correlate with disease severity, new cardiovascular events, new DUs and overall poor prognosis [1]. Microvascular abnormalities as assessed by nailfold videocapillaroscopy (NVC) and power Doppler ultrasound (PDUS) have been demonstrated to be predictive of new DUs occurrence [2,3].

Objectives: To study the severity of microvascular involvement at the 3rd and 4th finger of the dominant hand in patients with SSc with or without HDUs as assessed by NVC and 22-MHz PDUS.

Methods: 100 SSc consecutive patients fulfilling the 2013 EULAR classification criteria were enrolled. PDUS was performed at the 3rd and 4th finger of the dominant hand after exclusion of ulnar artery occlusion (UAO). In case of UAO non-dominant hand was examined. Ultrasound investigation was performed with Esaote MyLab 70 XVG by means of linear array transducer (10–22 MHz). Power Doppler settings were standardized (Doppler frequency 14.3 MHz, Gain 55%, PRF 750 Hz). PDUS measurements included sagittal scan of nailbed and fingertip qualitatively graded from 1 (no signal) to 4 (marked hyperemia) [4], and resistivity index (RI) of ulnar and radial proper digital arteries. Capillary density (number/mm) was calculated by NVC with magnification 200X performed on two images of the same digits examined by PDUS.

Results: 100 SSc patients, 87 (87%) women, 86 (86%) limited cutaneous SSc, median age 62.2 years old, median disease duration 8 years were evaluated. 7 (7%) patients had UAO. 33 (33%) had HDUs among them 23 had experienced more than one DUs and 2 had active DUs at the moment of evaluation. Semiquantitative perfusion score of sagittal scan of nailbed and fingertip were not significantly associated with the presence of HDU. RI and capillary density were significantly different in the two groups as shown in the table below:

	Presence of HDUs	Absence of HDUs	Mean difference (95% CI)
Capillary density	3.023	3.884	-0.862 (-1.390, -0.333)
RI	0.740	0.792	-0.052 (-0.090, -0.014)

Conclusions: A significant lower RI of ulnar and radial proper digital arteries reported in patient with HDUs is novel. By contrast PDUS grading of nailfold and fingertip were not significantly different in patients with or without HDUs. The finding of a significant lower capillary number assessed by NVC in patients with HDUs is consistent with previous results [2]. Adequately longitudinal studies exploring the predictive value of PDUS parameters are required to fully ascertain its role in SSc.

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SAT0365 MUSCULOSKELETAL US AND MRI FINDINGS IN JUVENILE SCLERODERMA

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Background: Musculoskeletal (MS) involvement and clinically evident arthritis occurs in up to 65% of patients with Juvenile Systemic or localized Scleroderma (JSc). It may be the first manifestation preceding even the onset of Raynaud or skin manifestations; patients presenting with arthritis, tenosynovitis or enthesitis may suffer from Sc. On the other hand clinical examination often underestimates MS involvement in Sc. US and MRI can help distinguish arthritis with effusion from dry tenosynovitis of Sc, define whether loss of range of motion (LOM) derives only from skin thickening or from bone and joint involvement and monitor disease progression.

Objectives: To describe the spectrum of MRI and US features in juvenile scleroderma with musculoskeletal involvement.

Methods: We describe MRI and color Doppler MSUS findings of clinically affected (with arthritis and/or LOM and/or overlying skin with edema or sclerosis) lower or upper extremities from 4 males and 2 females; 2 with systemic sc (ssc), 2 with linear scleroderma, 1 with generalized morphea, 1 with mixed morphea (median age 8.5 years, range 7–10.5; median time from symptom onset to MRI 11 months, range 2–24). MRI sequences: T1, fluid-sensitive, and T1-FS contrast-enhanced. Comparisons were made to uninvolved areas of the extremity, and the contralateral extremity. Findings guiding diagnosis and evaluation of disease extent are depicted in the Figure.

Results: Thickening of the dermis and infiltration of the subcutaneous fat with increase in signal intensity on fluid sensitive sequences and contrast-enhanced T1w images and hypointense signal lesions on unenhanced T1w images (asterisks) was apparent in 4 patients. In 2 male patients with generalized scleroderma, clinical LOM of fingers and wrists preceded skin sclerosis by 2 months. Joint and tendon sheath synovitis, indicated by initial MSUS, was detected in fluid sensitive and T1w enhanced images (white arrows). The combination of tendon-sheath synovitis and muscular fascia thickening and enhancement (black arrows), and contractures (thin white arrow) very characteristic of scleroderma,

helped identify sclerodermatous musculoskeletal involvement in the absence of skin induration. Focal bone marrow edema depicted as high signal intensity in fluid sensitive sequences (curved arrow) was found in 2 cases; 1 with generalized morphea without apparent overlying skin sclerodermatous lesion, 1 with linear scleroderma with atrophic lesions in all overlying structures

Conclusions: Musculoskeletal imaging features of juvenile scleroderma involving the skin, fascia, musculature and bones reflect pathomorphologic changes of this rare disorder and enable a complete assessment of the disease extent, including depth of infiltration and disease activity. In the described cases implementation of MSUS and MRI in Juvenile scleroderma led to earlier definition of the diagnosis and assisted the evaluation of disease extension.

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SAT0366 SMOKING BEHAVIOUR AND THE PROGRESSION OF ORGAN MANIFESTATIONS IN SYSTEMIC SCLEROSIS: A LONGITUDINAL EUROPEAN SCLERODERMA TRIALS AND RESEARCH GROUP STUDY

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Background: Systemic sclerosis (SSc) is a rare, multisystem autoimmune disorder. It is characterised by generalized microangiopathy, in which hypoxia and oxidative stress have been implicated in its pathogenesis. Tobacco inhalation increases free radicals and strongly promotes vascular damage. So far, data available with regards to a role of tobacco exposure with SSc severity and progression are scarce.

Objectives: We aimed to assess the effects of smoking on the speed of worsening of organ manifestations, namely lung involvement (forced vital capacity, FVC; forced expiratory volume, FEV1/FVC ratio; diffusing capacity for carbon monoxide corrected for alveolar volume, DLCO/VA), skin involvement (modified Rodnan skin score; mRSS), and digital ulcers (DU) in the European scleroderma trials and research (EUSTAR) database.

Methods: Adult SSc patients from the EUSTAR cohort with a follow-up visit 12±4 months after their baseline visit and available data on their smoking habits were included.

The associations of smoking behaviour (never smokers vs ex-smokers vs current smokers) with the disease manifestations at follow up were assessed after adjusting for potentially confounding covariates using multivariable linear or logistic regression analyses.

Missing data were imputed using multiple imputations.

Results: Of the 3,023 patients included (mean age 57 years, SD 13; 85% female), 66% stated that they never smoked, 23% were ex-smokers and 11% were current smokers. On average, ex-smokers had smoked for 19.5 years (SD 12) while current smokers smoked for 29.1 years (SD 13). Ex-smokers had smoked on average 17.3 pack-years (SD 20) and current smokers 29.3 pack-years (SD 36). The mean time since smoking cessation in ex-smokers was 15.8 years (SD 13). The FEV1/FVC ratio changed from 96.3 (SD 14) at baseline to 96.2 (SD 13) at follow up. Taking into account the effect of age, sex, autoantibody status, disease duration and SSc subset as well as the baseline values of the outcome, the FEV1/FVC ratio decreased faster in currently smoking SSc patients than in never smokers ($\beta = -2.8$, $p = 0.001$). Similarly, the DLCO/VA diminished faster in current

smokers ($\beta=3.7$, $p=0.006$) while on average the DLCO/VA decreased from 78.1 (SD 18) to 76.5 (SD 17). Changes in FVC were however not associated with smoking behaviour ($p=0.13$).

The mRSS changed between baseline and follow-up from an average of 7.6 (SD 7) to 7.3 (SD 7); on average, in smokers the mRSS improved faster than in never smokers ($\beta=-0.65$, $p=0.015$). The presence of DUs at follow-up was not associated with current smoking ($p=0.9$).

Having previously smoked was not significantly associated with the progression of any of the assessed outcomes.

Conclusions: Although the adverse effects of smoking on bronchial airways that is known in the general population could be replicated in the SSc population, the lack of a measurable adverse effect of smoking on SSc organ manifestations argues against a major role of tobacco associated free radicals and vasoconstriction in the pathogenesis of SSc vasculopathy and fibrosis.

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SAT0367 CAPILLAROSCOPY PATTERNS AMONG SYSTEMIC SCLEROSIS PATIENTS WITH GASTROINTESTINAL INVOLVEMENT AND MALNUTRITION EVALUATED BY SELF-REPORTED QUESTIONNAIRES

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Background: Gastrointestinal system (GI) is commonly involved in systemic sclerosis (SSc) beginning after early stages of disease.

Objectives: We aimed to investigate capillaroscopic findings among the SSc patients with GI involvement and malnutrition.

Methods: GI involvement was evaluated by UCLA SCTC GIT 2.0 questionnaire (Khanna D.) (7 multi-item; reflux, distension, soilage, diarrhea, social functioning, emotional wellbeing and constipation) in SSc patients ACR/EULAR classification criteria (2013). To report the nutrition status of the patients "malnutrition universal screening tool (MUST)" (sum of the scores of body mass index, weight loss in last 3–6 months and acute disease effect) scores were calculated. Simultaneously, nail fold video-capillaroscopy (NVC) was performed in all patients to determine early, active late scleroderma patterns (Cutolo et al.).

Results: In 58 SSc patients (51 female); the mean age, duration of Raynaud's, non-Raynaud symptom (year) and follow-up (month) were 46 ± 13 , 54 ± 50 , 10 ± 9 and 7 ± 7 , respectively. Limited cutaneous form, positive ANA and anti-Scl70, telangiectases, dysphagia, digital ulcers, lung disease were found in 40 (71%), 48 (83%) and 21 (36%), 45 (78%), 39 (67%), 33 (57%), 23 (40%) in patients, respectively. Of the NVC patterns, early was found in 16 (28%), active was in 11 (19%), late was in 28 (48%) and normal in 3 (5%). The scores of disease activity, severity and GI were shown to be higher in patients with late NVC pattern (table-1). Of the patients with MUST score of >0 ($n=10$), 9 had late and 1 had early NVC pattern.

Table 1. The scores of disease activity and severity and UCLA SCTC GIT 2.0 in SSc patients

	NVC (early)	NVC (active)	NVC (late)	p
Modified Rodnan skin score	4,3 $\pm$ 5,1	8,3 $\pm$ 6,9	12,3 $\pm$ 7,5	P=0,001*
Telangiectasia score (Shah)	2,3 $\pm$ 2,4	4,6 $\pm$ 1,9	6 $\pm$ 3,5	P<0,001*
Disease activity score (Valentini)	0,9 $\pm$ 0,9	1,1 $\pm$ 1,2	1,9 $\pm$ 1,2	P=0,014*
Disease severity score (Medsgger)	3,4 $\pm$ 2	4,6 $\pm$ 3	6,5 $\pm$ 2,6	P=0,001*
UCLA SCTC GIT 2.0	0,063 $\pm$ 0,111	0,259 $\pm$ 0,288	0,353 $\pm$ 0,327	P=0,002*
Reflux	0,263 $\pm$ 0,415	0,500 $\pm$ 0,509	0,874 $\pm$ 0,667	P=0,002*
Distension	0,118 $\pm$ 0,357	0,296 $\pm$ 0,498	0,564 $\pm$ 0,881	NS
Soilage	0 $\pm$ 0	0 $\pm$ 0	0,107 $\pm$ 0,315	NS
Diarrhea	0 $\pm$ 0	0,500 $\pm$ 0,741	0,268 $\pm$ 0,552	NS
Social functioning	0 $\pm$ 0	0,105 $\pm$ 0,237	0,184 $\pm$ 0,406	NS
Emotional wellbeing	0,011 $\pm$ 0,050	0,211 $\pm$ 0,372	0,118 $\pm$ 0,308	NS
Constipation	0,184 $\pm$ 0,380	0,159 $\pm$ 0,322	0,259 $\pm$ 0,469	NS

*Bonferroni, NS = not significant.

Conclusions: This SSc cohort predominantly had limited cutaneous disease and vascular manifestations. In patients with late scleroderma pattern, severity of GI was higher similar to disease activity and severity. Undernutrition was frequently seen in patients with late pattern. NVC might be useful to predict the severity of GI and malnutrition and allow to direct the patients to required procedures earlier.

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SAT0368 THE PREVALENCE OF OSTEOMYELITIS IN INFECTED DIGITAL ULCERS IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Skin ulcers, particularly digital ulcers occur in at least 50% of systemic sclerosis (SSc) patients (pts) and cause significant morbidity. They are often complicated by local infection which can lead to contiguous osteomyelitis.

Objectives: Our aims were to evaluate the accuracy of clinical diagnosis of osteomyelitis and to assess whether there are clinical parameters that may improve the precision of the clinical diagnosis.

Methods: We retrospectively analyzed the clinical data of consecutive SSc patients hospitalized for skin ulcers in a tertiary referral center for SSc. Our cohort is part of the EUSTAR cohort. The patients were evaluated by rheumatologists skilled in managing SSc skin lesions. All the patients with infected ulcers and suspected for contiguous involvement of underlying bone had bone scans. All the positive scans were followed with 99m-Technetium-Tc-labeled white blood cells (WBC) scintigraphy, in order to differentiate true osteomyelitis from acro-osteolysis or soft tissue infection. We collected demographic data, disease type, extent and severity, routine lab data (CBC, C-reactive protein -CRP, erythrocyte sedimentation rate (ESR), alkaline phosphatase (ALKP), albumin) and wound culture. Each hospitalization was considered a separate event. Statistical analysis: descriptive, student's T test, Mann-Whitney test.

Results: During the years 2003–2016, 220 SSc pts with skin/digital ulcers were hospitalized in our department for ilomedin treatment (993 hospitalizations). Most of the pts were hospitalized several times due to recurrent ulcers. Tc bone scan was performed in 39 pts (59 admissions) with infected ulcers (32 females, mean (SD) age 48 (15), disease duration 9 (6.6) years, 25 with diffuse SSc, skin score (MRSS) 9.9 (8)). Osteomyelitis was confirmed in 18 pts on 23 occasions. Osteomyelitis occurred twice in 5 pts in different locations. No statistically significant differences were found between the group with positive scans and the group with negative scans regarding demographic, clinical and lab data. 9 pts had 25 admissions for infected ulcers, osteomyelitis was confirmed in 14 of the admissions. No statistically significant differences were found for CBC ESR CRP ALKP between the osteomyelitis events and superficial infected ulcer admissions in these pts. The causative infectious agents were similar between the 2 groups. Positive bone and WBC scans confirmed osteomyelitis in 39% of clinical suspected cases. WBC scans confirmed osteomyelitis in 75% or the patients with positive Tc bone scans.

Conclusions: The prevalence of osteomyelitis among our SSc pts admitted for digital ulcers was 10%. The prevalence of confirmed osteomyelitis by scintigraphy in clinically highly suspected cases was 39%. Even when contiguous osteomyelitis was suspected by highly skilled rheumatologists, bone scan ruled out the diagnosis in 61% of the cases, thus avoiding unnecessary prolonged antibiotic therapy. No clinical predictors to rule osteomyelitis in or out could be identified.

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SAT0369 HETEROGENEITY IN THE UNDERLINING PATHOGENESIS IN PATIENTS WITH CONNECTIVE TISSUE DISEASE-ASSOCIATED BORDERLINE MEAN PULMONARY ARTERIAL PRESSURE AND ITS DISTINCTIVE HEMODYNAMIC CHARACTERISTICS FROM THOSE WITH NORMAL PULMONARY ARTERIAL PRESSURE

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Background: Borderline mean pulmonary artery pressure (PAP) (21–24 mmHg) may comprise a transition phase from normal pulmonary hemodynamic condition to pulmonary arterial hypertension (PAH), which is one of the fatal complications in connective tissue diseases (CTD) [1]. The accumulated evidence is not enough to conclude that treatment for PAH in this stage may be associated with improving clinical outcomes. On the other hand, CTD-associated PH is caused by various cardiopulmonary comorbidities. It is possible that borderline mean PAP, like overt PH, is caused not only by pulmonary vasculopathy but also by cardiac and pulmonary complications [2].

Objectives: To clarify whether patients with borderline mean PAP associated with CTD have distinctive hemodynamic characteristics from those with normal mean PAP and its underlining pathogenesis is heterogeneous as those with manifest PH.

Methods: We retrospectively investigated 76 CTD patients who underwent right heart catheterization from 2008 through 2016. Of the 76 patients, 25 (33%) and 16 (21%) had manifest PH and borderline mean PAP, respectively. The rest of the 35 patients had normal mean PAP. Systemic sclerosis was the most common