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 assed 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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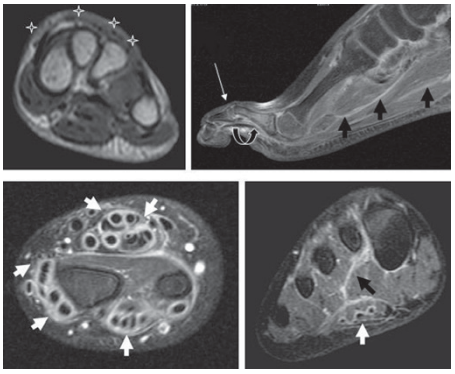
Background: Musculoskeletal (MS) involvement and clinically evident arthritis occurs in up to 65% of patients with Juvenile Systemic or localized Scleroderma (JSc). It may be the first manifestation preceding even the onset of Raynaud or skin manifestations; patients presenting with arthritis, tenosynovitis or enthesistis 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 (SD13).

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 (β=-2.8, p=0.001). Similarly, the DLCO/VA diminished faster in current