thickening +Raynaud's Phenomenon or peripheral vascular involvement +Positive ANAs+One visceral organ involvement typical of SSc +Absence of another defined connective tissue disease. Demographic, clinical and laboratory profile of all patients were analysed.

Results: Among the 249 patients included in this study, patients with limited cutaneous systemic sclerosis were 59.4% (n=148) and diffuse cutaneous systemic sclerosis were 31.3% (n=78). 9.2% (n=23) of patients satisfied poormoghim criteria for Systemic Sclerosis sine scleroderma. The following characteristics of Systemic Sclerosis sine scleroderma were observed in our cohort. Dyspnoea (82.6%) was the most common presenting complaint and Raynauds (100%) the most common sign observed. Most common internal organ involvement was oesophageal dysfunction (78%) followed by lung involvement in the form of interstitial lung disease (52.1%) and pulmonary hypertension (47.8%). Renal involvement was not observed in any of the patients. Most common autoantibody detected among patients with Systemic Sclerosis sine scleroderma was anti-centromere antibody (78.2%) followed by anti-scl70 antibody (21.8%). Nine patients (39.1%) received pulse cyclophosphamide therapy and 3 patients received rituximab therapy in view of interstitial lung disease.

Abstract AB0736 – Figure 1

Conclusions: The most common pattern of Systemic Sclerosis sine scleroderma observed in our cohort was Oesophageal dysfunction +Raynauds Phenomenon +Anti centromere antibody positivity +Interstitial lung disease with or without pulmonary hypertension which are usually the characteristics of limited cutaneous type of Systemic Sclerosis. Hence Systemic Sclerosis sine scleroderma could be a subset of limited cutaneous type without the phenotypic skin involvement.

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AB0737

AMINAPHTONE INCREASES SKIN BLOOD PERFUSION AND IMPROVES CLINICAL SYMPTOMS IN PATIENTS WITH RAYNAUD'S PHENOMENON INDEPENDENTLY FROM DIFFERENT TREATMENT BACKGROUNDS

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Background: Aminaphtone is a vasoactive drug that was recently demonstrated to improve both peripheral blood perfusion (BP) and clinical symptoms of Raynaud's phenomenon (RP) in patients with either primary or secondary RP to systemic sclerosis (SSc).¹⁻²

Objectives: The aim of this study was to evaluate possible interferences of different treatment backgrounds on both skin BP and RP-related clinical symptoms in patients treated with aminaphtone, during a six-month follow-up.

Methods: Forty-six patients with active RP were enrolled during routine clinical assessment after informed consent (11 primary RP, mean age 49±19 SD years, mean RP duration 6±3 years; and 35 secondary RP to systemic sclerosis, mean age 61±17 years, mean RP duration 11±9 years). Aminaphtone was orally administered 75 mg twice daily in addition to current treatments, and all patients were on a stable drug regimen for at least two months, which remained unmodified during the follow-up. All patients were taking cardioaspirin. Six groups of treatment backgrounds were identified: 1) no further treatments (12 patients); 2) hydroxy-chloroquine (2 patients); 3) colchicine (5 patients); 4) methotrexate (3 patients); 5) cyclosporine A (6 patients); 6) mycophenolate (6 patients); 7) proton-pump inhibitors (12 patients). Blood perfusion was measured by Laser Speckle Contrast Analysis (LASCA)³ at the level of fingertip, periungual areas, dorsum and palm of hands, and face at baseline (T0), after one (T1), four (T4), twelve (T12) and twenty-four (T24) weeks of treatment. Raynaud's condition score (RCS) and both frequency and duration of Raynaud's attacks were assessed at the same time. Statistical analysis was performed by non-parametric tests.

Results: During aminaphtone treatment, a progressive statistically significant increase of blood perfusion, as well as an improvement of RP clinical symptoms (decrease of RCS, frequency and duration of RP attacks/day), were observed in all above reported seven groups of RP patients with different treatments backgrounds from T0 to T12 in all skin areas (p<0.01). There were no statistically significant difference between the seven groups of patients concerning skin BP at different times (p=0.60). The results were similar in both primary and secondary (SSc) RP patients (p=0.40). Aminaphtone administration had to be stopped in 2 patients due to headache, and one patient was lost during follow-up.

Conclusions: This study demonstrates that the increase of skin blood perfusion and the improvement of RP clinical symptoms is not influenced by different treatment backgrounds in RP patients treated with aminaphtone. These preliminary results should be further confirmed by a randomised blind clinical trial.

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AB0738

IMPROVEMENT OF DERMAL THICKNESS IN SYSTEMIC SCLEROSIS PATIENTS TREATED WITH ENDOTHELIN RECEPTOR ANTAGONIST: LONG TERM STUDY BY HIGH FREQUENCY ULTRASOUND

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Background: Systemic sclerosis (SSc) is a connective tissue disorder characterised by skin involvement, which may be evaluated by both modified Rodnan skin score (mRSS) and skin high frequency ultrasound (US).¹⁻⁴ Endothelin-1 (ET-1) seems implicated in the development of dermal fibrosis in SSc.⁵ Bosentan, a dual ET-1 receptor antagonist seems effective in reducing skin fibrosis in SSc patients.^{6,7}

Objectives: The aim of this study was to evaluate by US the long-term effects of bosentan on dermal thickness (DT) in SSc patients, in combination with long-term cyclic intravenous iloprost versus iloprost monotherapy.

Methods: Thirty-eight SSc patients were enrolled during their standard treatment for digital ischemia. At baseline (T0), 19 patients already receiving cyclic intravenous infusion of iloprost (5 continuous days, average 80 mcg/day, every two

months), continued the treatment for further 4 years (T4) (ILO group). Other 19 patients, although they continued the same cyclic intravenous iloprost treatment as the previous group, also received bosentan 125 mg twice a day for 4 years (ILO+BOS group), due to digital ulcers. DT was yearly evaluated by both US (18 MHz probe, MyLab 25, Esaote, Italy) and mRSS at the level of the usual seven-teen skin areas (zygoma, fingers, dorsum of hands, forearms, upper-arms, chest, abdomen, thighs, legs, and feet).¹⁻⁴ Non-parametric tests were used for the statistical analysis.

Results: A statistically significant decrease of DT, measured by US, was observed in the ILO+BOS group from T0 (median DT 1.135 mm) to T4 (median DT 1.088 mm) ($p=0.01$). No statistical significant variation of mRSS was observed during the follow-up in this group of patients (median mRSS at T0 12/51 and at T4 11/51, $p=0.70$). Conversely, in ILO group, a statistically significant increase of DT was observed after four years, as measured by US (median DT at T0 1.070 and at T4 1.258, $p<0.0001$), as assessed by mRSS (median mRSS at T0 4/51 and at T4 8/51, $p<0.0001$). Transient increase of transaminases was managed by temporary bosentan discontinuation.

Conclusions: In this open study, the long-term treatment with ET-1 receptor antagonist in combination with iloprost seems to be associated with a decrease of DT in SSc patients, in contrast to the treatment with iloprost alone. DT evaluated by US over long term seems to be more susceptible to change than by mRSS.

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AB0739

LONGITUDINAL ASSESSMENT OF NAILFOLD CAPILLARY NUMBER, PERIPHERAL BLOOD PERFUSION AND DERMAL THICKNESS IN SYSTEMIC SCLEROSIS PATIENTS OVER A PERIOD OF 5 YEARS

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Background: Several studies demonstrated the relationship between blood perfusion (BP), dermal thickness (DT) and nailfold microangiopathy ("Early", "Active" and "Late" videocapillaroscopic patterns) in patients with systemic sclerosis (SSc).¹⁻³

Objectives: The aim of this study was to evaluate the changes of BP and DT, during a follow-up of 5 years, in SSc patients with persistent "Late" NVC pattern at baseline, as well as to confirm the correlations among microvascular damage extent, absolute nailfold capillary number (CN), BP and DT, during the follow-up.

Methods: Twenty-three female patients affected by SSc according to the LeRoy criteria⁴ (mean age 63±4 SD years, mean disease duration 6±4 years), displaying the most advanced "Late" NVC pattern of microangiopathy at baseline (T0), were enrolled and followed for five years (T5), after informed consent. Laser speckled contrast analysis (LASCA), skin high frequency ultrasound (US), modified Rodnan skin score (mRSS), and nailfold videocapillaroscopy (NVC) were yearly performed. Blood perfusion (BP), assessed by LASCA at the level of fingertips, periungual areas, dorsum and palm of both hands, was calculated as perfusion units (PU).¹⁻³ Dermal thickness (DT) was assessed by both US and mRSS in the same above reported areas.⁵ The microangiopathy evolution score (MES) and the CN per linear millimetre at first distal row were evaluated by NVC.^{6,7} Patients were receiving a wide range of drugs, including vasodilators, immunosuppressive agents and endothelin receptor antagonists. Statistical analysis was performed by non parametric tests.

Results: A progressive statistically significant decrease of both BP ($p<0.0001$) and nailfold CN ($p<0.0001$) values was observed from T0 to T5 at the level of all areas, as well as a progressive statistically significant increase of DT ($p<0.0001$), mRSS ($p<0.0001$) and MES ($p=0.01$) values. The progressive decrease of BP positively correlated over time with the worsening of nailfold CN ($p=0.03$, $r=0.62$), MES ($p=0.05$, $r=0.62$), mRSS ($p=0.002$, $r=0.72$) and DT ($p=0.002$, $r=0.64$).

Conclusions: Microvascular damage with progressive reduction of nailfold capillary number was found, in a five-year follow-up, associated with progressive

functional microvascular damage and DT worsening in the present cohort of SSc patients showing a persistent "Late" NVC pattern.

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AB0740

EVALUATION OF HAND DERMAL THICKNESS AND PERIPHERAL BLOOD PERFUSION IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Systemic sclerosis (SSc) is characterised by progressive skin involvement. The modified Rodnan Skin Score (mRSS) is the gold standard to assess skin damage, but it has significant limitations. Recently, several studies demonstrated the utility of skin high frequency ultrasound (US) as an alternative.

Objectives: The aim of this study was to identify possible correlations between peripheral blood perfusion (BP) and ultrasound dermal thickness (US-DT) at level of hand and finger in SSc patients.

Methods: Sixty-seven patients, satisfying the 2013 ACR/EULAR SSc criteria (mean age 64±9 SD years, mean disease duration 6±4 SD years) were enrolled. BP was measured as perfusion units (PU) by laser speckle contrast analysis (LASCA) at the level of dorsal region of hands. In a second time, different regions of interest (ROIs) were created at level of dorsum of the middle phalanx of 3rd finger and dorsum of hand bilaterally and the average BP was scored as perfusion units (PU), as previously reported.^{1,2} Both skin high frequency US and mRSS were used to evaluate DT at the above mentioned skin sites.² The same examinations were performed in 65 healthy subjects.

Results: BP was negatively correlated with both US-DT ($p=0.0005$) and mRSS ($p=0.007$) in SSc patients at the finger sites, but not at the level of dorsum of the hands. In healthy subjects, there was no statistically significant correlation between BP and DT as evaluated by both US and mRSS at either skin sites. SSc patients showed a statistically significant lower BP at the finger sites than healthy subjects ($p<0.0001$). No statistically significant difference in BP values was observed between SSc and healthy subjects at the dorsum of hand.

Conclusions: This study demonstrates a negative correlation between BP, as evaluated by LASCA and DT, as evaluated by both US and mRSS, in the 3rd finger of SSc patients. Additionally, the results confirm a reduced finger BP in SSc patients when compared to healthy subjects.

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AB0741

T-REG AND TH17 LEVELS IN PATIENTS WITH SYSTEMIC SCLERODERMA

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Background: Regulatory T-cells (T-reg) may play an inhibitory role in the development of autoimmune diseases (AID) by suppressing the immune response to autoantigens. Impaired or decreased T-reg and/or increased Th17 cells may be responsible for the development of AID. However, studies about the association of T-reg and systemic scleroderma (SSc) are limited and conflicting.

Objectives: Our aim is to determine whether there is a relationship between T-reg and Th-17 levels and disease activation in patients with SSc.