duration of follow-up was 15 months (range 3-59). At the end of the follow-up period, 10 of the 13 patients (77%) were still in treatment. In 2 patients (15%) the therapy failed, requiring lung transplantation in 1 case (after 7 months of therapy), while an autologous hematopoietic stem cell transplant was conducted in the other patient (after 13 months of anti-fibrotic). In one patient (8%) the treatment was discontinued due to adverse effects.

When quantifying the degree of response in patients who continued treatment, only 9 of the 10 patients were included in the analysis, since 1 had only 3 months of therapy. In these 9 patients there was an improvement of % pFVC (mean: +5.13; SD: 8.38) and % pDLCO (+4.42; SD: 5.06). The evolution of the efficacy variables analyzed is shown in the table.

The response in the Pulmonary function testing (PFT) in these 9 patients categorized according to the definitions of the American Thoracic Society was as follows: A) % pFVC: stabilization in 5 cases and improvement in 4; B) % pDLCO: stabilization in 7 cases and improvement in 3.

The frequency of adverse effects was 31% (4/13), but only the treatment was withdrawn in 1 patient (8%). Three patients presented digestive discomfort that forced to reduce the dose of the antifibrotic. One patient developed a toxic hepatitis due to nintedanib after 4 months of treatment that forced the withdrawal of the drug, replacing it 1 month ago with pifedronide. None of the patients died during the follow-up period.

Conclusion: Our data provides evidence of the safety and tolerability of both pifedronide and nintedanib in patients with DIDL-SAD refractory to GC and immunosuppressant therapy.

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SAT0257

THE SIGNIFICANCE ON THE TRANSITION THROUGH DIFFERENT PATTERNS OF NAILFOLD MICROVASCULAR DAMAGE ON THE COURSE OF SYSTEMIC SCLEROSIS

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Objective: To investigate the timing of transition through different patterns of nailfold microvascular damage in systemic sclerosis (SSc) patients and to determine the significance of this transition.

Methods: In the period January 2012 to December 2017, 400 SSc patients were followed up by nailfold videocapillaroscopy (NVC) the same operator (SPD) every 6 months. Demographic, clinical and laboratory data were recorded. The capillaroscopic findings were classified as normal, nonspecific or scleroderma pattern ("early", "active", or "late"). The evolution on the NVC pattern over time was monitored and recorded.

Results: The transition of microvascular damage through different NVC patterns of microangiopathy was seen in 53/400 (13.25%) SSc patients. At the baseline 11/53 (21%) patients had the non-specific changes, the early pattern had 25/53 (47%) patients, and the active pattern had 17/53 (32%) SSc patients. The mean $\pm$ SD time of progression from one to another NVC patterns was 27.68 ± 35.11 months. Improvement of the NVC patterns was found in 15/53 (28%) patients and deterioration in the remaining 38 (72%) patients. The mean time of progression from non-specific to early pattern (in 10 patients) and non-specific to active pattern (in 1 patient) was 14.5 ± 9.73 vs. 12 months, respectively. Time of changed from early to nonspecific (in 7 patients), early to active (in 17 patients) and early to late pattern (in 1 patient) was 30.85 ± 22.56 vs 22.29 ± 16.61 vs 24 months, respectively. Time of changed from active to early (in 8 patients) and active to late pattern (in 9 patients), was 26.25 ± 11.1 vs 18.00 ± 13.97 months, respectively. Progression of non-specific to early or active NVC pattern was related with limited form of SSc (91%), diffuse hand swelling (54%), arthralgia/arthritis (45%) and involvement of lungs (48%). Also, progression of early to active NVC pattern was related with limited form of SSc (64%), sclerodactyly (48%) and involvement of lungs (48%). In contrast, progression of active to late NVC pattern was related with diffuse form of SSc (53%), digital ulcerations (35%) and more frequent involvement of lungs (65%). These differences in the frequency of involvement of certain organs were not statistically significant.

Conclusion: These results demonstrate dynamic transition of microvascular damage through different NVC patterns of microangiopathy in about 13% of SSc patients. Patients with rapid progression from the early to active, as well as active to the late NVC patterns (<2 years) should be

monitored closely, because they are at risk of further significant damage to the internal organs.

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SAT0258

SEXUAL HEALTH IMPAIRMENT IN WOMEN WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is a chronic autoimmune disease that may affect all aspects of life including sexual function.

Objectives: To assess sexual function, pelvic floor function and sexual quality of life women with SSc compared to age-/sex-matched healthy controls (HC). To subanalyze sexual function in sexually active individuals.

Methods: In total 47 women with SSc (mean age: 50.5, disease duration: 5.6 years, lcSSc/dcSSc: 27/20, mRSS: 12.7, ESSG activity index: 2.5), who fulfilled the ACR/EULAR 2013 criteria, and 47 healthy women (mean age: 50.5) filled in 11 well-established and validated questionnaires assessing sexual function, pelvic floor function, quality of life, fatigue, physical activity and depression. Data are presented as mean $\pm$ SEM.

Results: Compared to HC, patients with SSc had significantly higher prevalence and greater severity of sexual impairment (FSFI, BISF-W), pelvic floor dysfunction (PISQ-12, PFIQ7), and worse sexual quality of life (SQoL-F) (table 1). There were no significant differences in sexual function between limited and diffuse SSc. Even sexually active SSc patients reported significantly greater sexual health impairment compared to sexually active HC. Sexual health impairment in SSc was associated with dis-

Table 1.

Questionnaire: score range	SSc (n=47)	HC (n=47)	p-value	SSc SA (n=29)	HC SA (n=29)	p-value
FSFI :2(worst)-36 (best)	14.7 $\pm$ 1.6	25.7 $\pm$ 1.6	p<0.0001	20.6 $\pm$ 1.7	28.5 $\pm$ 1.4	p<0.0001
BISF-W:-16(worst)-75(best)	16.6 $\pm$ 2.5	31.0 $\pm$ 2.6	p=0.0002	25.7 $\pm$ 2.8	34.9 $\pm$ 2.3	p=0.0143
SQoL-F: 0(worst)-100(best)	53.5 $\pm$ 3.7	79.9 $\pm$ 3.1	p<0.0001	58.0 $\pm$ 5.0	84.5 $\pm$ 2.6	p<0.0001
PISQ-12: 0(best)-48(worst)	14.6 $\pm$ 0.9	8.4 $\pm$ 0.6	p<0.0001	13.6 $\pm$ 1.0	7.9 $\pm$ 0.7	p<0.0001
PFIQ7: 0(best)-300 (worst)	29.0 $\pm$ 6.2	7.9 $\pm$ 2.0	p=0.0075	15.5 $\pm$ 5.0	9.3 $\pm$ 2.8	ns

Abbreviations: FSFI: Female Sexual Function Index; BISF-W: Brief Index of Sexual Function for Women; SQoL-F: Sexual Quality of Life Questionnaire; PISQ-12: Pelvic Organ Prolapse/Urinary Incontinence Sexual Questionnaire short form; PFIQ7: Pelvic Floor Distress Inventory Questionnaire, SA: sexually active; ns: not significant

Table 2.

Parameter: score range	Correlated parameters	r	p-value
FIS (fatigue): 0(best)-160(worst)	FSFI (sexual function)	-0.446	p=0.0016
MAF (fatigue): 1(best)-50(worst)	BISF-W (sexual function)	-0.445	p=0.0016
	SQoL-F (sexual quality of life)	-0.415	p=0.0050
BDI-II (depression): 0(best)-63(worst)	FSFI	-0.485	p=0.0005
	BISF-W	-0.483	p=0.0005
	SQoL-F	-0.375	p=0.0119
	PIFQ-7 (pelvic floor function)	0.445	p=0.0017
HAP (physical activity): 0(worst)-94(best)	FSFI	0.412	p=0.0040
	BISF-W	0.398	p=0.0055
SHAQ (scleroderma health status): 0(best)-100(worst)	FSFI	-0.557	p<0.0001
	BISF-W	-0.505	p=0.0002
	PIFQ-7	0.429	p=0.0025
ESSG activity index: 0(best)-10 (worst)	PISQ-12 (pelvic floor function)	0.322	p=0.0400
Disease duration	PIFQ-7	0.361	p=0.0124

Abbreviations: FIS: Fatigue Impact Scale; MAF: Multidimensional Assessment of Fatigue; BDI-II: Beck's Depression Inventory II; HAP: Human Activity Profile; SHAQ: Scleroderma Health Assessment Questionnaire; ESSG: European Scleroderma Study Group

ease activity/duration, health status, physical activity, fatigue and depression (table 2).

Conclusion: Women with SSc reported significantly impaired sexual function, sexual quality of life and pelvic floor function than age-matched HC. Worse scores in SSc were associated with disease activity/duration/health status, physical activity, fatigue, and depression.

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SAT0259

PREDICTIVE FACTORS FOR TREATMENT RELATED MORTALITY AND MAJOR ADVERSE EVENTS AFTER AUTOLOGOUS HEMATOPOIETIC STEM CELL TRANSPLANTATION FOR SYSTEMIC SCLEROSIS: RESULTS OF A LONG TERM FOLLOW-UP MULTI-CENTRE STUDY

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Background: Autologous hematopoietic stem cell transplantation (HSCT) has shown superior efficacy to cyclophosphamide pulse therapy in systemic sclerosis (SSc) but its application is hampered by high treatment related mortality (TRM). To date factors, other than smoking, predicting TRM are unknown.

Objectives: To describe event-free survival and TRM after HSCT and to evaluate the predictive value of baseline characteristics. Event-free survival was defined as the absence of mortality or major organ failure (i.e. cardiac failure (left ventricular ejection fraction (LVEF) < 30%, respiratory failure (resting PaO₂ < 8kPa/60 mm Hg or PaCO₂ > 6,7 kPa or chronic oxygen supplementation), or renal failure requiring dialysis).

Methods: All patients who started with the procedure of HSCT for SSc in the Netherlands between 1.1.1998 and 31.12.2016 were included. Except in 4 patients, eligibility criteria and procedures of the ASTIS trial were used (1). A Kaplan-Meier event-free survival curve was constructed. All deceased patients were discussed in a consensus meeting, to determine the cause of death as TRM, progression of SSc or other. The univariate association of TRM and event-free survival with baseline characteristics (gender, age, disease duration, smoking ever, FVC, DLCO, LVEF) was examined by Cox regression analysis.

Results: 92 patients were included, including 4 patients with limited cutaneous subtype who were treated because of interstitial lung disease. The median (IQR) follow-up time was 4.5 (2.3 – 12.3) years. Twenty patients died during the observation period of whom 10 (10.8%, 9 males) in the first year because of TRM. Twelve patients (13.1%) developed major organ failure (6 lung, 3 kidney and 3 heart) of whom 8 died during the

Table 1.: demographic and disease related factors

Male/female	49/43
LcSSc/DcSSc	4/88
Age at HSCT mean (SD) (year)	46.8 (10.3)
Disease duration at HSCT mean (SD)(year)	2.4 (2.4) median 1.6
mRSS mean (SD)	26 (10.1)
Antibodies: ANA, ATA, Anti-RNP positive	88/92; 34/92; 1/92
Ever smoker	54/84
FVC as% pred. mean (SD)	84.9 (22.7)
DLCO as% pred mean (SD)	55.1 (16.2)
LVEF < 50% (by echo)	3/92

observation period. Event-free survival after five and ten years was 68% (40/59 patients) and 59% (30/51), respectively. TRM was significantly associated with male gender (HR (95% CI): 8.7 (1.1 -68.9), but not with smoking; events were associated with male gender, older age and LVEF <50% at baseline (HR: 4.08 (1.5 – 11.0); 1.1 (1.0-1.1) and 4.7 (1.1-20.6), respectively).

Conclusion: In this national cohort study of HCST in patients with SSc and poor prognosis we observed a median event-free survival of 4.3 years. TRM was 10.8% and major events occurred in 13%. TRM was correlated to male gender. Low LVEF, male gender and older age were correlated with major events.

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SAT0260

MUSCULAR INVOLVEMENT OF THE LOWER LIMBS IN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOPATHIES: A MRI EVALUATION

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Background: Since many years, muscular magnetic resonance imaging (MRI) has been used in the diagnosis and follow-up of patients with idiopathic inflammatory myopathies (IIM), but the clinical significance and the pattern of the muscular alterations (edema, fatty infiltration and atrophy) in different subsets of the disease are still not well defined.

Objectives: The aims of this study were to analyse possible differences in the involved muscles between dermatomyositis (DM) and polymyositis (PM); the correlations between the pattern of muscular involvement in different subsets of the disease and disease parameters in a monocentric cohort of patients.

Methods: We retrospectively collected data from 85 patients with poly and dermatomyositis (EULAR/ACR criteria) who performed pelvic and thighs muscle MRI from January 2010 to December 2018: 27 had DM and 58 PM, mean age 58.6±13.4 years, mean disease duration of 45±73 months. The images in 22 muscles for the presence of muscular edema, fatty infiltrates and atrophy were assessed by a dedicated radiologist. Moreover, data about serum creatine kinase (CK) and manual muscle