Table 2. McNemars test comparing PASP estimated by repeat TTE to hemodynamic parameters measured by repeat RHC when PVR is also considered

Repeat TTE	Repeat RHC	
	Stable or improved PASP and PVR	Deteriorated PASP or PVR (>15%) or PVR (>15%)
Stable or improved PASP	24	10
Deteriorated PASP (>15%)	5	11

P=NS (0.3).

can be safely monitored in the long-term by TTE. Further studies to help identify those patients at need for follow-up RHC are warranted.
Disclosure of Interest: None declared
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FRI0415 CAPILLAROSCOPIC ABNORMALITIES CORRELATE WITH ORGAN DAMAGE IN CHINESE PATIENTS WITH SYSTEMIC SCLEROSIS

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Objectives: To study the nailfold capillaroscopic patterns and microangiopathy evolution score (MES) and correlate with the severity of organ damage in Chinese patients with systemic sclerosis (SSc)
Methods: Patients who fulfilled the 2013 ACR criteria for SSc were studied. A full physical examination was performed. Blood was taken for SSc autoantibodies, along with a full lung function test and echocardiogram. The extent of skin involvement was assessed by the modified Rodnan skin score (mRSS). Organ damage of SSc was assessed by the Medsger disease severity scale. A Nailfold capillaroscopic examination was performed by a trained nurse blinded to the medical history of the patients. The following parameters were obtained: (1) capillaroscopy patterns (early, active and late); (2) Degree of enlarged capillaries, giant capillaries, capillary haemorrhages, capillary density, disorganization of vascular array and capillary ramification assessed by a semi-quantitative method; and (3) MES score (sum of capillary density, disorganization of vascular array and capillary ramification). Correlation among the capillaroscopic patterns, individual capillaroscopic parameters and the MES with organ damage was performed by the Spearman's rank correlation test.
Results: A total of 138 Chinese patients were studied (91.3% women; age 56.36±11.81 years). The median disease duration was 8.14±6.21 years. 39 (28.3%) patients had DcSSc and 99 (71.7%) had LcSSc. Anti-centromere, anti-Scl 70 and anti-RNA polymerase III antibodies were present in 28.6%, 28.5% and 5.6% of the patients respectively. Organ damage was present in all patients, most common being skin (84%), lung (79%), peripheral vascular (74%) and GI tract (46%). The median mRSS was 6 (IQR 2–12). A total of 27 patients (19.7%) had early SSc pattern on capillaroscopy, 40 (29.2%) had active pattern and 68 (49.6%) had late pattern. The median MES score was 3.02 (IQR 1.76–5.25). Patients with late SSc pattern on capillaroscopy had significantly longer disease duration and were more likely to have organ damage in the general, peripheral vascular and lung domains compared to those not having late SSc patterns. The total MES score correlated significantly with organ damage scores in the muscle (Rho 0.188; p=0.029), GI tract (Rho 0.169; p=0.048) and lung (Rho 0.265; p=0.006) domains. Regarding individual components of the MES score, capillary density correlated significantly with scores in the peripheral vascular (Rho 0.460; p<0.001), skin (Rho 0.343; p<0.001), joint/tendon (Rho 0.220; p=0.011), muscle (Rho 0.295; p=0.001), GI tract (Rho 0.188; p=0.028) and lung (Rho 0.238; p=0.015) damage domains. Enlarged capillaries correlated significantly with scores in the muscles (Rho -0.205; p=0.017) and lung (Rho -0.213; p=0.029) damage domains. Giant capillaries and microhaemorrhages correlated significantly with scores in the peripheral vascular (Rho 0.239; p=0.005 and Rho 0.228; p=0.007 respectively) damage domains. Disorganization of capillary array correlated significantly with scores in the lung (Rho 0.253; p=0.009) damage domain. Capillary ramifications correlated significantly with the scores in the kidney (Rho 0.171; p=0.048) damage domains.
Conclusions: In Chinese patients with SSc, capillaroscopic patterns and components of the microangiopathy evolution score were associated with severity of organ damage.
Disclosure of Interest: None declared
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FRI0416 CHARACTERISTICS AND CAPILLAROSCOPIC FINDINGS OF SYSTEMIC SCLEROSIS PATIENTS WITH SEVERE PERIPHERAL VASCULAR INVOLVEMENT RECEIVING SPECIFIC VASODILATOR TREATMENT

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Background: Severity of peripheral vascular involvement (PVI)is known as an important determinig cause of morbidity in sytemic sclerosis (SSc). Different vasodilating agents have been found to reduce severity and contribute to healing of digital ulcers (DU)in SSc.
Objectives: We aimed to evaluate the characteristics and capillaroscopic patterns of the patients with severe PVI under different vasodilator therapeutic regimens.
Methods: The patients were grouped as "severe PVI" if score of PVI is ≥2 (PVT=2,digital pitting scars; PVT=3, digital tip ulcerations and PVT=4, digital gangrene; Medsger) and "non-severe PVI" if score of PVI is ≤1 (PVI=0,no Raynaud's; PVI=1, Raynaud's requiring vasodilators). We included patients with severe PVI who received cyclic iloprost and bosentan and/or sildenafil and compared to non-severe PVI. Nail fold video-capillaroscopy (NVC)was assessed qualitatively (Cutalo et al., early, active and late patterns).
Results: Severe PVI group more frequently had diffuse cutaneous form, contractures, lung disease, anti-Scl70 positivity and high acute phase response and exposed to immunosuppressives (table-1).

Table 1. Demographics and characteristics of SSc Patients

	Non-severe PVI n=32	Severe PVI group 2 n=22	p
Age (year)/female (%)	44±15/ 88%	48±12/ 87%	NS
Duration of Raynaud's/nonRaynaud's (year)	9±9/5±5	12±10/9±9	NS
Diffuse Cutaneous involvement	5 (16%)	10 (45%)	P=0,039
Anti-Scl70 (+)	7 (22%)	11 (50%)	P=0,024
Interstitial lung disease	7 (22%)	14 (64%)	P=0,001
Flexion contractures	3 (9%)	10 (45%)	P=0,008
Echo PAP>40 mmHg	2 (6%)	4 (18%)	NS
High acute phase response	12 (38%)	18 (82%)	P=0,002
FVC ≤80%/	3 (9%)	12 (55%)	P<0,001
DLCO ≤80%	10 (31%)	14 (64%)	P=0,011
Immunosuppressives- CYC (pulse)	5 (16%)	9 (41%)	P=0,018
– Azathioprin	8 (25%)	10 (46%)	P=0,030
– Mycphenolate mofetil	2 (6%)	10 (46%)	P<0,001
– Rituksimab	1 (3%)	6 (27%)	P=0,027
– Steroids (PRD<10 mg/d)	11 (34%)	17 (77%)	P=0,001

NS = not significant.

Scores of telangiectasia, skin, activity and severity were lower in non-severe group. NVC late pattern was frequent and early pattern was rare in severe PVI groups. Forty-one% (9/22) of patients received second oral agent for PVI. Monotherapy and combination groups had similar scores (table-2).

Table-2: Capillaroscopy Patterns among SSc groups

	non-severe PVI n=32	severe PVI n=22	Cyclic iloprost+ bosentan n=10	Cyclic iloprost+ bosentan+sildenafil n=9
NVC -normal	3(9%)	0	0	0
-early pattern	12(38%)**	2(9%)	1(10%)	1(11%)
-active pattern	10(31%)	2(9%)	2(20%)	0(0%)
-late pattern	8(25%)	18(82%)*	7(70%)	8(89%)
Telangiectasia score (Shah)	3,7±2,5**	6±3,7	4,8±3,2	7,1±4,2
Modified Rodnan skin score	5,8±5*	13,5±8,7	13,6±9,3	13,2±7,6
Activity score (Valentini)	0,9±0,9*	2,1±1,2	2,2±1,3	2,2±1,2
Severity score (Medsger)	3,5±1,7*	7,5±2,4	7,8±2,3	7,4±2,7

**p<0,001, **p<0,05 when severe and non-severe PVI compared*

Conclusions: NVC late pattern was frequent in severe PVI group with long disease duration, exposed to intensive immunosuppresives and received specific vasodilators for PVI. Disease activity and severity were higher in severe PVI group. One third of the severe group required oral combination therapy for PVI. Monotherapy and combination groups were similar in terms of severity. NVC is a useful method to monitor digital ischemia and severe organ involvement in SSc.
Disclosure of Interest: None declared
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FRI0417 EXERCISE-INDUCED PULMONARY HYPERTENSION IN SYSTEMIC SCLEROSIS PATIENTS: TRANSCRIPTOME ANALYSIS OF PERIPHERAL BLOOD AT THE EARLY STAGE OF THE DISEASE

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Background: Pulmonary arterial hypertension (PAH) is prominent as a vascular involvement in systemic sclerosis (SSc), which remains a leading cause of death in spite of current best treatments. As the pulmonary vascular disease (PVD) can be well compensated for, more than a half of the pulmonary circulation is impaired before early PAH is detected. Although recent studies focused on molecular basis of the PVD, the underlying mechanisms have not been fully elucidated, especially at the early stage of the disease.
Objectives: To detect the subclinical PVD, and to explore the changes in transcriptome of peripheral blood at the early stage of SSc associated PAH.
Methods: Total of 74 cases without PAH symptoms (NYHA I) with either Raynaud phenomenon (RP: n=61), skin sclerosis (n=43) or SSc-related autoantibody (anti-RNP: n=10, centromere: n=36, topoisomerase-1: n=2, RNA polymerase III: n=2) were enrolled. To detect the latent PAH, exercise Doppler echocardiography (DE) with Master's two-step stress was carried out. Systolic PAP (sPAP) was determined by maximum velocities of tricuspid regurgitation jets, and exercise induced pulmonary hypertension (exPH) group was segregated from normal response group (exN) with using the definition of a sPAP greater than 40 mm Hg during exercise, or a exercise increase in sPAP by greater than 20 mm

Hg[1)]. For transcriptome analysis, total RNAs from whole peripheral blood cells were extracted with using PAXgene miRNA kit. After constructing single-stranded, strand-specific libraries, multiplex sequencing was done. After quantifying the expressions of transcripts, differentially expressed genes (DEGs) between exPH and exN group were selected by paired T-test ($P<0.05$). And then, hierarchical clustering analysis and pathway enrichment analysis (PathVisio) were performed.
Results: There were no significant differences between exPH and exN group in the result of total skin score, serum BNP, tests of pulmonary function and thermography after 0℃-stress. Positive SSc-related autoantibody was a risk factor for exPH (odds ratio, 1.41); especially, positive anti-RNP seemed to be prominent (odds ratio, 3.21). Based on the 817 DEGs between exPH and exN group, the hierarchical clustering showed major 4 clusters, and one of them consisted of only cases in exPH group. When we focused on 117 genes reported to be directly implicated in the development of PAH[2)], it is noteworthy that 4 of them including TGF-β induced protein were differentially expressed. Pathway analysis of transcriptome revealed that 22 pathways, such as hypertrophy model, lung fibrosis and Wnt/B-catenin signaling, were differently enriched between exPH and exN group.
Conclusions: The paradigm of SSc-PAH management should ideally be aimed at detecting early PVD and starting treatment prior to fulfilling the criteria for PAH. Although detection of early PVD in SSc patients remains a major challenge, exercise DE seemed to be a good, non-invasive method for screening. It is noteworthy that expression changes in some of known PAH-related genes were detected from peripheral blood of exPH patients. It shows the possibility that the therapeutic intervention at early stage of the disease may alter the clinical course.
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FRI0418 FACTORS ASSOCIATED WITH STEROID-FREE REMISSION IN PATIENTS WITH INFLAMMATORY MYOPATHIES. A RETROSPECTIVE ANALYSIS OF A SINGLE-CENTER COHORT

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Background: The inflammatory myopathies are a heterogeneous group of connective tissue diseases characterized by muscle weakness and inflammation. Corticosteroids are the standard main treatment for inflammatory myopathies. However, steroid therapy often causes a wide range of side effects. Although immunosuppressive drugs are used as steroid-sparing agents in an effort to prevent disease recurrence, the appropriate duration of steroid use remains unclear.
Objectives: We investigated whether steroid therapy can be safely withdrawn in patients with inflammatory myopathies followed in a single center.
Methods: We retrospectively reviewed clinical charts of 71 consecutive patients (age 51.9±15.7 y.o., female 69%) who met Bohan and Peter criteria for polymyositis (PM)/dermatomyositis (DM) and modified Sontheimer's criteria for clinically amyopathic dermatomyositis (ADM), respectively. Steroid free remission was defined as a 3-month consecutive period of no disease activity without corticosteroid treatment. Factors associated with steroid free remission were examined.
Results: Of 71 identified patients, 29 patients (40%) were DM, 15 patients (21%) were PM, 9 patients (13%) were overlap myositis, and 18 patients (25%) were ADM. Thirty-seven patients (52%) had muscle weakness, 5 patients (7%) had malignancies and 43 patients (61%) had signs of interstitial lung disease. With a mean follow-up of 6.6±5.0 years, 9 patients (13%) died during follow-up period. The remaining 62 patients were treated with corticosteroids alone or in combination with immunosuppressants. Steroid-free remission was achieved in 21 of 62 patients (34%) patients with a mean time to steroid withdrawal of 5.5±4.0 years. Six of 21 patients (29%) relapsed 1.7±1.7 years after steroid withdrawal. There were no differences in onset of age, disease duration, positive ANA, positive Anti Jo-1 antibodies, serum creatine kinase levels, maximum dose of corticosteroids, skin, joint and lung involvement between steroid-free group and non-steroid-free group. Elevated inflammatiory markers were associated with long-term steroid use (p=0.038). Concominant immunosuppressants were more frequently used in non-steroid-free group than steroid-free group (p=0.002).
Conclusions: Steroid-free remission might be achieved in some patients with inflammatory myopathies.
Disclosure of Interest: None declared
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FRI0419 THE PREDICTIVE PROGNOSTIC FACTORS FOR CLINICAL COURSE OF POLYMYOSITIS/DERMATOMYOSITIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

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Background: Interstitial lung disease (ILD) and concomitant infectious diseases are the predominant causes of death in polymyositis/dermatomyositis (PM/DM). We have already reported that hypocapnea and ILD lesion in upper lung fields are independent prognostic factors. Micro RNA is a non-coding RNA, which has a certain function such as transcriptional regulation. miR-1 has been reported to be associated with myocyte differentiation and to decrease in muscle tissue from patients with inflammatory myopathies.
Objectives: Here we investigated the association of serum miR-1 level with clinical course of PM/DM-associated ILD (PM/DM-ILD).
Methods: We retrospectively analyzed clinical baseline, serum miR-1 level, initial therapeutic regimens, total amounts of PSL, clinical outcomes, and episode of infection of patient with PM/DM-ILD who had received initial treatment at six hospitals associated with Yokohama City University from 2003 to 2016. The serum miR-1 level was measured by quantitative real-time PCR.
Results: One hundred sixteen (PM 22, DM 51, and clinically amyopathic DM 43) patients were included. The mean age was 56±15 years and 83 were female. As initial therapies, oral PSL, methylprednisolone (mPSL) pulse, intravenous cyclophosphamide (IVCY), and oral calcineurin inhibitor therapies were performed in 113 (97%), 80 (69%), 48 (41%) and 80 (69%), respectively. Forty-one patients had a serious infection at 51±38 days from initiation of immunosuppressants and 10 died of infections. Old age, low $PaCO_2$ and albumin, high LDH and KL-6, high score of ILD, high initial dose of PSL, mPSL pulse, IVCY, calcineurin inhibitor and combination therapy were extracted as risk factors for infection by univariate analyses. A multivariate logistic regression analyses revealed that combination therapy (p=0.012, OR 2.83), old age (p=0.024, OR 2.12), high initial dose of PSL (p=0.024, OR 2.69), low albumin (p=0.031, OR 3.56), and low $PaCO_2$ (p=0.038, OR 2.67) were independent risk factors for infection. Serum samples were obtained from total of 14 patients and 13 healthy controls. Serum miR-1 levels in PM/DM-ILD patients before treatment were significantly higher than those in healthy controls (p=0.047). Also serum miR-1 levels were significantly higher in PM/DM-ILD patients with concomitant infectious diseases as compared to patients without infectious diseases (p=0.043). We further divided the PM/DM-ILD cases into two groups by the serum miR-1 level. The higher miR-1 group showed poorer effectiveness of ILD treatment (p=0.040), and lower lymphocyte count (p=0.013) as compared to the lower miR-1 group.
Conclusions: Appropriate monitoring is important for PM/DM-ILD, especially in older patients with malnutrition or decreased respiratory function. miR-1 can be a new biomarker for predicting treatment response and concomitant infectious diseases during treatment for PM/DM-ILD.
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FRIDAY, 16 JUNE 2017

Spondyloarthritis - etiology, pathogenesis and animal models

FRI0420 ASSOCIATION OF SUPRESSOR OF CYTOKINE SIGNALING -3 (SOCS-3) EXPRESSION WITH INTERLEUKIN-23 RECEPTOR (IL-23R) SINGLE NUCLEOTIDE POLYMORPHISMS (SNPS) IN ANKYLOSING SPONDYLITIS (AS)

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Background: Nowadays genetic-association studies have discovered new genes, other than *HLA-B27*, as *IL-23R* associated with AS. The signalling pathway trough IL-23R is negatively regulated by the SOCS proteins. However, the reports regarding the roles of SOCS in AS are very rare at present.[1,2]
Objectives: The aim of this study is to assess the gene expression of *SOCS-1*,