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.
**References:**
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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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# 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*,