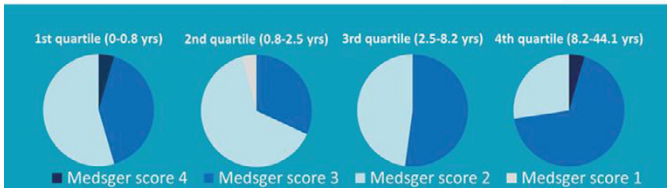
Results: Of 422 SSc patients in the database, 344 patients had SSc meeting ACR criteria. 89 patients were exclusively ATA-positive, of which n=42 with mild disease and n=47 with severe disease. Patients with severe disease appeared to be younger (mean age 50 vs 55 yr), more often non-caucasian (51 vs 12%), with a longer time since non-Raynaud (median 4 vs 2 yr) and more often diffuse skin involvement (dcSSc;62 vs 41%), calcinosis (6 vs 0%) and weight loss (23 vs 7%). Stratification by disease duration, however revealed there are no real differences between mild en severe disease. Overall 47% of ATA+ patients in our cohort presented with mild SSc. When stratifying patients according to time of non-Raynaud, 55% of ATA+ patients presented with mild disease in the first disease duration quartile (median follow-up 0.5 years, range 0–0.8 years). In the forth disease duration quartile, according non-Raynaud time (median follow-up 12.7 years, range 8.2–44.1) the percentage with mild disease was still 27% (Figure 1).

Figure 1. Medsger scores of ATA patients stratified by duration non-Raynaud



Conclusions: In our cohort, 47% of ATA-positive patients presented with mild systemic sclerosis, which could not be explained by disease duration. This suggests that solely the presence of ATA is of limited clinical relevance. Readily available sociodemographic and clinical parameters including type of skin involvement seem to have only limited value in identifying ATA patients with more severe SSc. More complex serological findings as antibody titers and fine-specifity of ATA should be defined for optimal serological subsetting.

Disclosure of Interest: None declared

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AB0651 THE EFFECT OF CYCLOPHOSPHAMIDE ON PULMONARY FUNCTION AND DEPENDENCE ON DISEASE ACTIVITY OF INTERSTITIAL LUNG DISEASE ASSOCIATED WITH SYSTEMIC SCLEROSIS

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Background: The pathogenesis of interstitial lung disease associated with systemic sclerosis (SSc-ILD) is not completely elucidated, although it is believed that chronic alveolar inflammation leads to increasing fibrosis. Treatment strategies using cyclophosphamide (CYC) have been focusing on the inflammatory pathway of SSc-ILD. We hypothesized that CYC is more effective in patients that are in the early, inflammatory phase.

Objectives: The objectives of this study are to analyze the effects of intravenously CYC pulses (750mg/m^2) on pulmonary function (FVC, DLCO) in SSc-ILD after 12, 24 and 36 months, and whether this effect is dependent on the extent of ILD, the proportion of ground glass compared to fibrosis, SSc disease duration or baseline DLCO <60%.

Methods: Patients with SSc-ILD receiving CYC pulses between 2003 and 2015 were classified by the Goh (2008) criteria in either limited or extensive ILD, using HRCT at baseline independently judged by two raters. Pulmonary function tests were performed at 0, 6, 12, 24 and 36 months. Missing outcome data due to drop-out were replaced by last observation carried forward, except in case of death.

Results: Seventy-five patients were included, 33 with limited ILD, 42 with extensive ILD. There were no baseline differences in age, gender, SSc subtype classification, disease duration or autoantibody status. Mean FVC and DLCO were stable after 12, 24 and 36 months of follow-up (figure 1).

There was no effect in the degree of change in mean FVC and DLCO for the different effect modifiers (table 2): the extent of ILD, proportion of ground glass compared to fibrosis, short SSc disease duration or baseline DLCO <60%.

Conclusions: Mean FVC and DLCO in SSc-related ILD was stable during 36 months of follow-up after cyclophosphamide pulse therapy. The extent of ILD, proportion of ground glass, SSc disease duration and baseline DLCO <60% did not influence the effect of CYC on pulmonary function. CYC did not appeare to be more effective in the early phase of SSc-ILD.

Disclosure of Interest: None declared

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AB0652 EARLY ORGAN INVOLVEMENT, SURVIVAL AND CAUSES OF DEATH IN AN UNSELECTED COHORT OF DUTCH PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) patients are at risk for organ involvement and premature death. The occurrence of organ involvement that is reported differs widely between various long term cohort studies; ILD 25–90%, PAH 8–32%, CI 5–30%, and SRC 4–12%. Differences in findings also apply to survival, the 5- and 10-year survival rates between studies vary from 80% to 90% and from 60% to 85% respectively (1–3).

Objectives: To assess the occurrence of organ involvement and death in a large, unselected cohort of Dutch SSc patients at the moment of diagnosis and during 5 years of follow-up, stratified by disease subtype and auto-antibodies.

Methods: Up to 2015, 690 SSc patients were included in the Nijmegen SSc cohort. Occurrence of interstitial lung disease (ILD), pulmonary arterial hypertension (PAH), cardiac involvement (CI), scleroderma renal crisis (SRC) and occurrence of death were determined using survival analysis, stratified by disease subtype (limited cutaneous SSc and diffuse cutaneous SSc) and auto-antibodies (ACA, ATA, anti-RNP).

Results: Organ involvement was already present at SSc diagnosis in 32% of patients. In 25%, organ involvement developed during follow-up, mostly ILD (22%). Significant differences between lcSSc and dcSSc were found in SRC at baseline and ILD, PAH and SRC during follow-up. Between the autoantibody subgroups, the occurrence of ILD, PAH and SRC at baseline and ILD during follow-up differed. There were no differences in survival between subtypes and auto-antibodies. The overall 5-year survival rate was 89%. Patients without organ involvement at SSc diagnosis had a better 5-year survival rate than patients with organ involvement at SSc diagnosis: 95% versus 73% respectively ($p<0.001$). (figure 1)

Conclusions: In many SSc patients, organ involvement is already present at diagnosis or develops in the first 5 years after diagnosis.

Survival is significantly worse in patients who already have involvement at the moment of SSc diagnosis.

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AB0653 EFFICACY OF AN INTENSIVE 24-WEEK PHYSIOTHERAPY PROGRAMME IN MYOSITIS PATIENTS - PRELIMINARY DATA FROM A SINGLE-CENTER CONTROLLED STUDY

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Background: Involvement of musculoskeletal system (inflammatation, atrophy and permanent damage to the muscle) in idiopathic inflammatory myopathies (IIM) leads to impaired function and reduced muscle strength, endurance, aerobic capacity and decreased quality of life. Data on efficacy of non-pharmacologic care in IIM is very limited due to variety in studied interventions/outcomes.

Objectives: To address the limitations of existing studies, and evaluate the effect of a controlled, long-term (24-week intervention, 24-week follow-up), intensive (1h physiotherapy twice weekly, and home-exercise for 1h 5x weekly), tailored physiotherapy program to improve muscle strength, endurance and deep stabilizer system, and quality of life/disability in cohorts with a substantial number of IIM patients.

Methods: All patients fulfilled the Bohan and Peter 1975 diagnostic criteria for dermatomyositis (DM) or polymyositis (PM), had skeletal muscle involvement, and were consecutively recruited from 2014 to 2016 at the Institute of Rheumatology in Prague. Both groups received educational materials and instructions for home exercise at baseline, however, only intervention group underwent the intensive physiotherapy programme. At months 0,3,6,12 all patients were assessed by a physician [physical examination, Myositis intention to treat index (MITAX), Myositis disease activity assessment visual analogue scale (MYOACT), and Myositis damage index (MDI)], and a physiotherapist blinded to intervention [standardized tests evaluating the level of muscle strength [Manual muscle test-8 (MMT-8)], and endurance [Functional index-2 (FI-2)], patients filled out patient reported outcomes (PRO)/questionnaires [HAQ, SF-36, Beck's depression inventory-II