months during the first year of treatment. Some of the patients received both drugs in different evolutionary periods of their disease. The study was approved by the Clinical Research Ethics Committee (CREC) of our hospital.
Results: Of the 42 patients included, 29 received Pi, 69% men and 31% women, with a mean age of 71 years (78% ex-smokers). Baseline FVC was 2140 ml (74.4% of the predicted value) and DLCO was 40.8% with respect to the expected value. The absolute loss in FVC after 52 weeks of follow-up was 200 ml. 48.3% required treatment with glucocorticoids (GC) at some point, either due to exacerbations of the disease or as concomitant treatment. 65.5% presented some adverse reaction to Pi, being gastrointestinal discomfort (GI) the most frequently observed, although mainly of self-limiting course, with the definitive suspension of the drug being necessary in 6 cases. As for the patients treated with Ni, 70.6% were men and 29.4% women, 82% ex-smokers, with an average age of 72 years. Baseline CVF value was 2480 ml (83.8% of the predicted value) and DLCO value was 54.7%. The decrease in FVC in absolute terms was 70 ml. Similarly, 4 patients required the use of GC at some point in the study. With regard to adverse reactions, 76.5% presented some type of adverse event, GI discomfort being the most frequent, followed by increased transaminases and mild diarrhoea. The great majority were of limited duration, requiring the definitive suspension of the drug in 5 patients. Five patients treated with Pi died due to exacerbations of their disease.
Conclusions: This project supports, with data from usual clinical practice, the beneficial effect of the AF drugs available for the treatment of mild-moderate IPF. Both drugs have been shown to slow down the natural evolution of the disease, reducing the loss of FVC, a variable directly related to mortality. This therapy has acceptable safety margins. However, there are still no references regarding its administration in incipient and advanced stages of the disease nor on their combined use with each other or with immunomodulators for the control of immune mediated diseases.
Acknowledgements: To the nurses and all members of the Pneumology Service for their collaboration in the follow-up of the patients included in this study.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.7208

FRI0462 SERUM KL-6 IS A STRONG PREDICTOR FOR RELAPSE OF MYOSITIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

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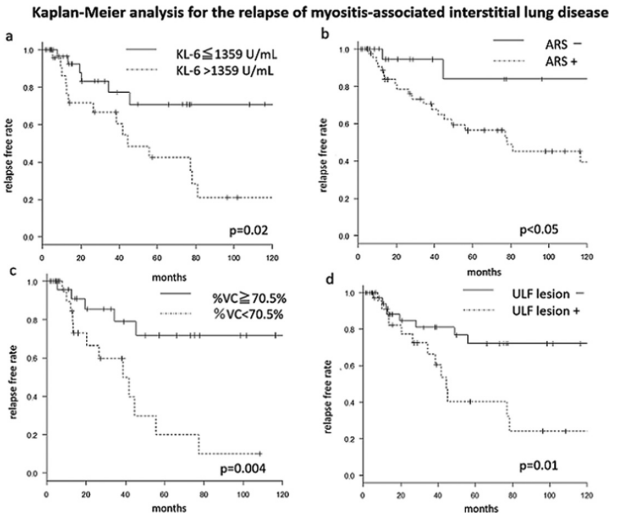
Background: Polymyositis (PM), dermatomyositis (DM) and clinically amyopathic DM (CADM) are autoimmune myositis which can be associated with interstitial lung disease (ILD).[1,2] The relapse rate of ILD is high, reported as approximately 20%–55%.[3,4] Since relapses result in decreased pulmonary function, it is important to identify the predictive factors for the relapse.
Objectives: The aim of this study was to elucidate the predictive factors for the relapse of ILD associated with myositis (PM/DM/CADM).
Methods: We conducted an observational retrospective study. Patients with myositis-associated ILD who have ever visited our institution between 2002–2017 and achieved remission once were enrolled. Patients who died before remission were excluded. We collected their clinical information from medical records. We compared patient characteristics between relapse group and non-relapse group by Fisher's exact test or Mann-Whitney U test at first. Relapse was defined as exacerbation of radiological findings of which doctor-in-charge decided to intensify therapy for ILD. We performed Kaplan-Meier analysis to compare the relapse-free survival for the characteristics that had significant differences between two groups. To perform Kaplan-Meier analysis, continuous variables were converted to dichotomous variables for analysis by setting cut-off values determined by receiver-operating characteristics (ROC). Then, using each characteristic which showed significant difference in Kaplan-Meier analysis, we conducted Cox's proportional hazard analysis for multivariate analysis. For relapse group, we examined the changes of serum KL-6 levels from the initial treatment of myositis-associated ILD to the relapse of ILD. We calculated the average of serum KL-6 levels of 3 months and 6 months before relapse, respectively, then compared them with KL-6 levels at the time of relapse.
Results: Seventy-two patients with myositis-associated ILD at our institution were enrolled. Among 72 patients, 24 experienced relapse (relapse group) and 48 did not experience relapse (non-relapse group). Median observational period was 31.5 months and 39.0 months, respectively. Median levels of serum KL-6, the rate of patients who had upper lung field (ULF) lesion by CT, and anti-ARS antibody prevalence were significantly higher in relapse group than in non-relapse group (1870 vs 935 U/mL, p=0.003; 62 vs 27%, p=0.01; 88 vs 60%, p=0.03, respectively). Median levels of%VC was significantly lower in relapse group than in non-relapse group (65.7 vs 81.2%, p=0.02). By ROC analysis, the cut-off levels of serum KL-6 and%VC were determined as 1359 U/mL and 70.5%, respectively. Kaplan-Meier analysis showed serum KL-6 >1359 U/mL(p=0.02), anti-ARS antibody (p<0.05),%VC <70.5 (p=0.004), and ULF lesions (p=0.01) were significantly related to the relapse (figure 1). Multivariate analysis revealed only serum KL-6 >1359 U/mL was an independent risk factor for relapse (hazard ratio: 4.9 (95%CI 1.0–24.0), p<0.05) among the 4 characteristics. At the time of the relapse, serum KL-6 levels were increased 37% from the 3 months average and 51% from the 6 months average.



Conclusions: Serum KL-6 was a strong predictor for relapse of myositis-associated ILD.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.2045

FRI0463 A NEW COMPUTED TOMOGRAPHY INDEX FOR QUANTIFICATION OF INTERSTITIAL LUNG DISEASE IN SYSTEMIC SCLEROSIS IS ASSOCIATED WITH LUNG FUNCTION PARAMETERS IN THE SHORT TERM FOLLOW-UP

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Background: New computer-assisted methods for the objective quantification of interstitial lung disease (ILD) at computed tomography (CT), based on the evaluation of mean lung attenuation (MLA), skewness and kurtosis have been recently investigated in Systemic Sclerosis (SSc). We developed a computerised integrated index (CII) based on a weighted evaluation of MLA, skewness and kurtosis and investigated its reliability for the quantitative assessment of SSc-ILD and its associations with lung function parameters in a cross-sectional study.
Objectives: To identify the CII cut off value with the highest sensitivity and specificity for CT-detected ILD and to investigate its impact on lung function parameters over-time of baseline assessed CII.
Methods: SSc patients meeting the new ACR/EULAR classification criteria, who had undergone a volumetric lung CT study from July 1 st 2014 to June 30th 2015, had been evaluated at baseline for ILD quantification by Goh et al. method and the previously referred dedicated software and had their CII calculated, were enrolled in a prospective study including complete clinical, serological, and functional assessment at baseline and at 1 year follow-up (FU).
Results: Thirty-nine out of 83 (47%) SSc patients (79 females, 4 males; mean age 56.4±11.3 years; median disease duration 12 years;[2–54] 18 diffuse cutaneous and 65 limited cutaneous SSc) had evidence of ILD as assessed by volumetric CT of the lungs at baseline. CII in patients with ILD was significantly lower than in those without ILD (−0.4929±0.9933 versus 0.4145±0.8059 HU; p<0.0001). ROC analysis revealed that the best discriminating CII value for ILD was 0.1966:

sensitivity 0.81 (95% C.I. 0.68 to 0.92); specificity 0.66 (95% C.I. 0.52 to 0.80). Out of the 44 ILD negative patients, 22 (50%) presented a CII value lower than the cut-off, and 13 of them (59%) were found to have a diffusing lung capacity for CO (DLCO)<80% of predicted. At 1 year FU, the CII was significantly correlated with total lung capacity -TLC (r=0.45, p=0.004) and DLCO (r=0.29, p=0.045). Out of the 22 patients with a CII <0.1966 but no ILD at visual evaluation, 11 (50%) developed a FVC decline at 1 year, and 8 (36.7%) a DLCO decline.
Conclusions: Here we confirm that quantitative computer-assisted CT of the lungs could be a reliable method for SSc-ILD evaluation and found that it could also be useful in predicting the evolution of lung function in the short-term FU.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.7559

FRI0464 CLINICAL MANIFESTATIONS OF PERSISTENT ANTIPHOSPHOLIPID ANTIBODIES IN SYSTEMIC SCLEROSIS PATIENTS

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Background: Antiphospholipid antibodies (aPL) can be detected in systemic sclerosis (SSc) patients. While their clinical signification is well known in the primary antiphospholipid syndrome (APS), their frequency and clinical association remain debated matter of issue. In a previous study, we reported the prevalence and their clinical association of a single aPL testing a French cohort of SSc patients. Yet, the persistence of aPL during follow-up of patients has never been assessed.
Objectives: This study aimed to assess i. the persistence of aPL at least 12 weeks after the first dosage and ii. the clinical associations.
Methods: At least 12 weeks apart after the first test, 213 SSc patients were consecutively tested for lupus anticoagulant (LA), anticardiolipin (aCL) and anti-ß2glycoproteine 1 (anti-ß2Gp1) antibodies. aCL and anti-β2GpI were measured using commercial ELISA assays (Orgentec, Trappes, France), positivity was defined as ≥10 UGPL/mL (aCL) or ≥10 UA/mL (anti-β2GpI). Clinical associations were studied using binomial logistic regressions.
Results: The mean time between two tests was 13.2±6.3 months. One or more type of aPL was persistent in 7 patients (3%). Patients with persistent aPL developed more frequently venous thrombosis (VT) [4/7 (57%) versus 23/205 (11%), p=0.006]. Arterial or/and venous thrombosis were more common in patients with persistent aPL than other patients [4/7 (57%) versus 33/205 (16%), p<0.001]. Persistent aPL was also associated with miscarriages [4/6 (67%) versus 33/155 (21%), p=0.023].

Association between aPL positivity persistance and clinical manifestations

	aPL positivity	aPL negativity	p
Arterial or venous thrombosis	4 (57.1%)	33 (16.1%)	**<0.001**
Venous thrombosis	4 (57.1%)	23 (11.2%)	**0.006**
Arterial thrombosis	1 (14.3%)	14 (6.8%)	0.406
Miscarriage	4 (66.7%)	32 (20.6%)	**0.023**
Digital ulceration	0 (0%)	40 (19.8%)	0.351
PAH	1 (14.3%)	13 (6.7%)	0.402

Conclusions: The prevalence of persistent aPL in our cohort was 3%. VT and miscarriages were associated with persistent aPL.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.6640

FRI0465 QUANTITATIVE ASSESSMENT OF INTERSTITIAL LUNG DISEASE IN IDIOPATHIC INFLAMMATORY MYOPATHIES: VISUAL SCORE AND COMPUTERISED PROCESSING ANALYSIS OF HIGH RESOLUTION COMPUTERISED TOMOGRAPHY

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Background: High resolution computed tomography (HRCT) is an essential technique for the characterisation of interstitial lung disease (ILD) in patients with idiopathic inflammatory myopathies (IIM). Although several visual scores are available for a semiquantitative evaluation of ILD, an automatic quantitative analysis of lung involvement may represent a valuable improvement to reproducibly determine the extent of the disease.
Objectives: To compare the semiquantitative visual score to a volumetric texture and local volumetric histogram feature-based analysis software (CALLIPER) to quantify the lung involvement in IIM patients.
Methods: 21 consecutive IIM patients (Bohan and Peter criteria) who underwent a HRCT between November 2016 and November 2017 were prospectively enrolled. Twelve were affected by polymyositis and 9 by dermatomyositis. We collected smoking habits data and myositis specific autoantibodies positivity, respiratory symptoms according to MRC dyspnea scale and two patients reported outcome questionnaires: Leicester cough questionnaire (LCQ) and St. George's Respiratory questionnaire (SGRQ). Patients underwent a non-contrast, supine, volumetric CT (≤2 mm slice thickness) that was evaluated by an expert radiologist according to Warrick's score (WS) and also quantified by CALLIPER (Mayo Biomedical Imaging Resource – USA). CALLIPER analysis of the extent of interstitial lung abnormalities (ground glass, lung reticulation, honeycombing) was performed and a total score, expressed as percentage of lung involved (ILD%), was obtained. Analysis of the vascular involvement were performed as percent of pulmonary vessel volume (VESS%).
Results: Between the 21 patients, five (23.8%) were smokers. Four patients were positive for anti-synthetase autoantibodies -ASS (3 Jo-1, 1 Pl-7), 1 SRP, 1 Mi2. Dyspnea was present in 19 patients (90%): 9 grade 1, 7 grade 2, 3 grade 3 according to MRC scale. There were strict correlations between WS and ILD (r=0.802 p<0.001) and VESS% (r=0.746 p<0.001). LCQ score was negative correlated to WS (r=−0.728 p=0.007) but the correlations were stricter with ILD score (r=−0.868 p<0.001) and VESS% (r=−0.801 p<0.002). SGRQ did not correlate both to WS and CALLIPER parameters. Patients with ASS have higher ILD and VESS% compared to seronegative patients (respectively p=0.003 and p=0.006).
Conclusions: CALLIPER represents an innovative technique to quantify the lung involvement in IIM patients: the main advantage of CALLIPER is the reproducibility that avoids the inter and intra-reader bias. Although more data are needed in larger cohorts, the use of CALLIPER may open new routes for the evaluation of ILD in IIM patients, both in medical practice and in randomised controlled trials.
Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2018-eular.4265

FRI0466 ARTICULAR AFFECTION IN SYSTEMIC SCLEROSIS: CORRELATION OF ULTRASOUND FINDINGS WITH CLINICAL, BIOLOGICAL AND RADIOGRAPHICAL FINDINGS

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Objectives: To evaluate the prevalence of subclinical synovitis in systemic sclerosis (SS), as well as the correlation of sonographic findings in hands with radiographs and with clinical-analytical parameters. In addition, the correlation with scales of functionality will be determined.
Methods: Cross-sectional observational-analytical study that included 40 patients with ES and 23 patients with rheumatoid arthritis (RA) as a control group during the period 10/2015–03/2016. Clinical, analytical, immunological, ultrasound and the radiological characteristics of both groups and their respective statistical correlations were compiled.
Results: The mean age of the group with SS was 53.3±14.1 years; 87.5% were women and 75% of the cases were cutaneously limited. The average time of evolution of the disease was 6.8 years from the diagnosis. Regarding the clinical joint involvement in the carpus and hands, frank arthritis was present in 16 cases (40%), with an average of DAN: 3.77 and TAN: 1.57; with an average of EVA global pain: 30/100 and elevated CRP in 42.5% of cases. The prevalence of subclinical synovitis with ultrasound expression of synovial effusion ±synovial hyperrtrophy in the SS group was at carpal level (77%), FCM (60%), PFI 32.5%) and DFI (15%). The statistical linear correlation between findings of cortical erosions at the ultrasound level versus the radiological data was positive only in the involvement at the joint level FCM and FID. In the group with SS, the HAQ questionnaire