systemic sclerosis. Altered level of albumin and decreased appetite may lead to worsening in nutritional status. Assessment of nutritional status in this group of patients should be performed regularly, because it can be potentially modified.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6032

SAT0328 AN OPEN-LABEL STUDY OF AMBRISENTAN WITH ANTI-FIBROTIC AGENT COMBINATION THERAPY IN THE TREATMENT OF DIFFUSE SYSTEMIC SCLEROSIS

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Background: Systemic sclerosis (SSc) is marked by immune dysregulation, inappropriate fibrosis and a vasculopathy for which there is currently no universally accepted disease modifying regimen. Ambrisentan, a selective type A endothelin receptor antagonist (ERA) has known benefits in the treatment of the vasculopathy related to pulmonary arterial hypertension and has been postulated to have anti-fibrotic effects. The additive effect of an ERA in combination with an anti-fibrotic agent has not previously been studied in SSc.
Objectives: To determine the safety and efficacy of ambrisentan in combination with an anti-fibrotic agent in early diffuse cutaneous systemic sclerosis (dcSSc).
Methods: Patients already on anti-fibrotic therapy for early dcSSc with onset of skin sclerosis less than 48 months before study entry were placed on ambrisentan 5mg daily for 12 months in an open-label study. Laboratory and clinical parameters to assess safety, as well as severity and progression of SSc were obtained at specified intervals. The primary outcome measure was the modified Rodnan skin score (mRSS), and secondary outcomes were the Medsger severity score, the Short Form Health Survey (SF)-36 questionnaire, pulmonary function tests (PFTs), and 2D echocardiograms (echo).
Results: A total of 15 patients were recruited who were on anti-fibrotic therapy upon entry of the study, most commonly mycophenolate (14 patients) and 1 patient on methotrexate. Of the patients entering the study, 10 patients (66.7%) completed 12 months of treatment with the study drug. Using intention-to-treat analysis, the mRSS improved significantly with a mean difference in mRSS of -8 from study entry to study end (p=0.000167). Among study completers (n=10), there was a trend for improvement in all but one category of the SF-36 while only the physical health component was of statistical significance (p=0.025). The median Medsger severity scores remained unchanged except for a change in median skin score. No statistically significant change was observed in PFTs and in mean estimated pulmonary arterial pressure by echo for those patients in whom data was obtained at baseline and at 12 months (n=12 and n=5, respectively). The most common adverse events included peripheral edema and dizziness. Two patients withdrew from the study due to intolerance of the study drug. Serious adverse events (SAE's) occurred in 4 study subjects and included scleroderma renal crisis (n=1), upper gastrointestinal bleeding (n=1) and infections (n=2). None of the SAEs were deemed to be related to the study drug.
Conclusions: In this prospective open-label study of ambrisentan and anti-fibrotic combination therapy in early dcSSc, we observed significant improvements in the mRSS and patient-perceived physical health status on the SF-36. There was no observed change in peripheral vascular involvement and no consistent trend in PFTs. Combination ambrisentan and anti-fibrotic therapy appears relatively safe in this 12-month study. Larger, controlled trials are needed to further investigate the safety and efficacy of combination therapy.
Acknowledgements: [C.T. Derk received funding support for this study by Gilead Pharmaceuticals]
Disclosure of Interest: A. Schorpion: None declared, M. Shenin: None declared, R. Neubauer: None declared, C. Derk Grant/research support from: Gilead Pharmaceuticals
DOI: 10.1136/annrheumdis-2017-eular.3932

SAT0329 INTRAVENOUS CYCLOPHOSPHAMIDE ACCORDING TO THE EURO-LUPUS NEPHRITIS PROTOCOL FOR PROGRESSIVE INTERSTITIAL LUNG DISEASE IN PATIENTS WITH POLYMYOSITIS/DERMATOMYOSITIS

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Background: Interstitial lung disease (ILD) affects 30–70% of patients with Polymyositis (PM) and Dermatomyositis (DM) and is one of the major contributors of morbidity and mortality.
Objectives: To study the efficacy and the safety of intravenous cyclophosphamide (IVCYC) according to the Euro-Lupus nephritis protocol for ILD in PM/DM patients.
Methods: Twelve patients with PM/DM (mean age 54± SD 8), who received, 500 mg IVCYC every other week, up to 12 times, according to the treating physician, as first line treatment, were retrospectively studied. Six patients had anti-Jo1, 4 anti-PL7, 1 anti-PL12, and 1 anti-MDA5 auto-antibodies. The median (IQR) disease duration before IVCYC was 4 (10,8) months. High doses of prednisolone were given for the first month and then gradually tapered. Response to treatment after a median (IQR) follow-up of 5 (2,8) months was based on pulmonary function tests (PFT) and HRCT). The extent of pure ground-glass opacity (pGGO), pulmonary fibrosis (PF), honeycomb cysts (HCs) and emphysema was scored (0=0%, 1=1–5%, 2=6–15%, 3=16–20%, 4=21–25%, 5=26–50%, 6=51–75%, 7= >75%) in the upper, middle and lower lung zones before and after therapy. The total score for each finding was calculated as the sum of the scores of the 3 zones (1).
Results: The mean IVCYC total amount was 4.75 ± SD 1.4 gr. Before IVCYC, the median (IQR) values of forced vital capacity (FVC)%, forced expiratory volume in 1 second (FEV1)%, vital capacity (VC)%, total lung capacity (TLC)% and diffusion capacity of the lung for carbon monoxide (DLCO)% were 67 (26), 60 (14), 63 (11), 63 (12) and 57 (25), respectively. After therapy, the median (IQR) values became 74 (29), 80 (18), 80 (24), 77 (19) and 68 (27), respectively. The difference between baseline and follow-up TLC%, FVC% (fig.1) and VC% median values was statistically significant ($p<0,05$). FVC% and TLC% improved >10% in 6 and 5 patients, respectively; DLCO% improved >15% in 3 patients. Before IVCYC, the median (IQR) scores of pGGO and PF were 12,5 (9) and 12 (7), respectively. After IVCYC, they decreased (7 (6) and 9 (12), respectively). In the group of anti-Jo-1 positive patients, the difference was close to the statistically significant (from 13,5 (10) to 7,5 (15), p=0,06 and from 9,5 (10) to 4,5 (16), p=0,07, respectively). The median (IQR) pGCO scores of anti-Jo-1 negative patients improved (from 11,5 (11) to 8,50 (17)), while the median (IQR) PF scores were unchanged (from 13,5 (4) to 14 (10)). At baseline and follow up, the median scores of HCs and emphysema were 0. No statistically significant correlations were found between PFT values and HRCT scores. The difference of PF extent was negatively correlated with the disease duration before the first IVCYC (r=-0,56, p=0,056, fig 1). No adverse events or drug toxicity were observed.
Conclusions: After IVCYC according to the Euro-Lupus nephritis protocol PFT and HRCT findings improved in PM/DM patients with ILD without any adverse events or drug toxicity. Longitudinal controlled studies are needed to confirm the efficacy and the safety of this treatment protocol.

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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.5878

SAT0330 LOWER PREVALENCE BUT COMPARABLE CLINICAL CHARACTERISTICS AND PROGNOSIS OF SYSTEMIC SCLEROSIS IN CRETE-GREECE AS COMPARED TO OTHER EUROPEAN COUNTRIES: A SINGLE CENTER EXPERIENCE

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Background: Systemic Sclerosis (SSc) is a rare, multisystemic connective tissue disease with significant morbidity. Prevalence and incidence of SSc varies worldwide (0.7–265 cases per 100,000/y[1,2] and 0.06–12.2 cases per 100,000/y[1] respectively). Mortality in SSc patients remains 3.5 times higher than the general population[3]. We sought to generate updated data on the epidemiology and burden of SSc in the Greek population.
Objectives: To study the prevalence and incidence of SSc, describe the clinical characteristics and assess mortality and main causes of death in Crete (Greece) over a 5-year period (2010–2015).
Methods: We conducted a cohort Study in defined geographic area (6,5% of Greek population). We reviewed demographics, clinical features, autoantibodies status and the causes of death from the Scleroderma cohort followed at the Rheumatology clinic of the University Hospital, Heraklion. The cause of death was categorized as related or not to scleroderma. Incidence and prevalence were estimated including patients living permanently in Crete who fulfilled SSc 2013 ACR/EULAR Classification Criteria for Scleroderma.
Results: 72 patients (88% women, mean±SD age at diagnosis 48.5±16.7 years [range 15–87]) were identified, corresponding to a crude point prevalence of $13.8/10^5$ (CI 95% 11–17/10^5) (December 2015). The Incidence rate was estimated at $0.05/10^5$ per year (period 2010–2015). Diffuse SSc (dSSc) was present in 27.7%, limited SSc (lSSc) in 72.2%, while an overlap syndrome in 19.4% (9.7% with systemic lupus erythematosus). Frequencies of anti-Scl70 and anti-cectromere antibodies were 59.7% and 9.7%, respectively. Arthritis was present in 69.5%, lung involvement in 61% (9.7% pulmonary arterial hypertension [PAH]), whereas only a single patient developed renal crisis. 8.3% developed cancer. Case fatality rate during 2010–2015 was 9.7% (CI 95% 7.4–11.9) with an average (±SD) age of death at 65.2 (± 17.6) years. Mortality cases were related to SSc in 30.7%. The main cause of death was sepsis (30.8%) followed by PAH and cardiac arrest (15.4% each). Male gender ($p<0.001$) and presence of PAH (p=0.001) were related to mortality. Mean disease duration until death was 5.3