and secondary dermatomyositis (DM) and polymyositis (PM) including the rest of the patients. Also, IIM were classified (II) as primary polymyositis (PPM), primary dermatomyositis (PDM), overlap syndrome (OSd), juvenile myopathies (JM), cancer-associated myopathies (CAM), autoimmune necrotizing myopathy and inclusion body myositis (these were grouped as other myositis; OM).

Results: In the PM and DM groups 250 and 229 (52.2% and 47.8%) patients were included respectively. Positive ANA, anti-Jo1 and anti-RNP were higher in the PM than in the DM group (67, 22 and 19% vs. 56, 11 and 6%) ($p=0.21$, $p=0.002$, $p=0.0001$, respectively). The presence of anti-MI2 was higher in the DM group ($p=0.024$), according to the classification II, we found statistically significant differences in ANA, anti-Jo1, anti-RNP and anti-ACA. The OSd group had the highest proportion of ANA, anti-RNP and ACA positive AA and the JM group had the lowest frequency of Anti-Jo1 (see Table 1).

Table 1. Autoantibodies profile in IIM according to classification II

AA	Comparison between groups		Frequency	P
ANA	OSd vs.	PPM	91% vs. 54%	0,0001
		PDM	91% vs. 63%	
		JM	91% vs. 40%	
		CAM	91% vs. 63%	
Anti-Jo1	JM vs.	JM	63% vs. 54%	0,0001
		OM	3% vs. 25%	
Anti-RNP	OSd vs.	PPM	3% vs. 25%	0,0001
		PDM	3% vs. 19%	
		JM	3% vs. 36%	
		OM	3% vs. 36%	
ACA	OSd vs.	PPM	40% vs. 6%	0,0001
		PDM	40% vs. 3%	
		JM	40% vs. 7%	
		CAM	40% vs. 5%	
ACA	OSd vs.	PDM	18% vs. 3%	0,002

Conclusions: The AA associated with the IIM subtypes is consistent with published scientific evidence on other cohorts for ANA, anti-Jo1, anti-RNP and anti-MI2, in spite of the small sample size. The OSd group showed higher ANA and anti-RNP frequencies which might be explained by the coexistence of SLE and MCTD patients. It could be interesting to follow up those PPM patients with positive AA because they could be in the future diagnosed with a connective tissue disease. Carrying out longitudinal studies that include a greater proportion of patients may help to evaluate and predict the clinical course of IIM.

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Acknowledgements:

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.7050

AB0626 EPIOTOPE PROFILING OF ANTI-RO52 ANTIBODIES IN PATIENTS WITH SYSTEMIC SCLEROSIS, SYSTEMIC SCLEROSIS-ASSOCIATED PRIMARY BILIARY CIRRHOSIS, AND PRIMARY BILIARY CIRRHOSIS ALONE

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Background: Anti-Ro52 antibodies are detected in patients with Sjogren's syndrome (SjS), systemic sclerosis (SSc) and other autoimmune rheumatic diseases. Epitope mapping studies of anti-Ro52 in autoimmune rheumatic diseases have failed to find any difference on epitope recognition suggesting a common stimulus for the loss of tolerance to Ro52. Such epitope differences, however, were noted between SjS and non-rheumatic diseases and in particular, SjS-associated primary biliary cirrhosis (SjS-PBC) or PBC alone (1).

Objectives: To assess whether or not the Ro52 epitope profile in anti-Ro52+ SSc patients differs from that of SSc-associated PBC (SSc-PBC) or PBC.

Methods: Serum samples were obtained from 63 anti-Ro52-positive (by ELISA) patients (33 SSc, 10 SSc-PBC, 20 PBC alone). Antibody reactivity to full length recombinant Ro52 and 5 baculovirus expressed Ro52 fragments spanning the whole sequence [Ro52-1 (aa 1–129), Ro52-2 (aa 125–268), Ro52-3 (aa 269–475), Ro52-4 (aa 57–180, partly overlapping with Ro52-1 and Ro52-2), and Ro52-5 (aa 181–320, partly overlapping with Ro52-2 and Ro52-3)] were tested by a line immunoassay.

Results: Reactivity was present to: full length Ro52 in all anti-Ro52 positive SSc, PBC-SSc, or PBC patients by line immunoassay; Ro52-1 in 6/33 (18.2%) SSc, 3/10 (30%) SSc-PBC, and 3/20 (15%) PBC; Ro52-2 in 33/33 (100%) SSc, 10/10 (100%) SSc-PBC and 20/20 (100%) PBC; Ro52-3 in 0/33 (0%) SSc, 0/10 (0%) SSc-PBC and 0/20 (0%) PBC; Ro52-4 in 4/33 (12.1%) SSc, 2/10 (20%) SSc-PBC, and 4/20 (20%) PBC and Ro52-5 in 11/33 (33.3%) SSc, 4/10 (40%) SSc-PBC, and 9/20 (45%) PBC respectively ($p>0.05$ for all epitopes amongst the 3 cohorts). No statistically significant correlation was found.

Conclusions: The major epitope of anti-Ro52 does not differ among SSc, PBC-SSc and PBC and is localized on aa125–268, the major epitopic region of anti-Ro52 antibodies in SjS and other rheumatic diseases. Our epitope mapping data suggest that the mechanisms responsible for the loss of tolerance to Ro52 is common amongst diseases.

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Disclosure of Interest: C. Liaskos: None declared, A. Gkoutzourelas: None declared, T. Simopoulou: None declared, E. Marou: None declared, T. Scheper Employee of: Euroimmun AG, W. Meyer Employee of: Euroimmun AG, L. Sakkas: None declared, D. Bogdanos: None declared

DOI: 10.1136/annrheumdis-2017-eular.5299

AB0627 RISE-SSC: A DOUBLE-BLIND, RANDOMISED STUDY EVALUATING THE EFFICACY AND SAFETY OF RIOCIQUAT FOR THE TREATMENT OF PATIENTS WITH DIFFUSE CUTANEOUS SYSTEMIC SCLEROSIS

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Background: Diffuse cutaneous systemic sclerosis (dcSSc) is a severe subtype of SSc, a rare, chronic autoimmune connective tissue disease associated with significant morbidity and mortality, and with reduced quality of life. Currently, there are no approved disease-modifying therapies to treat the vascular and fibrotic damage in patients with dcSSc. Soluble guanylate cyclase stimulators, such as riociguat, target the transforming growth factor beta pathway, inhibiting signalling through non-SMAD pathways. They have been shown to inhibit dermal fibrosis in several animal models and to ameliorate fibrosis of the skin and intestine in bleomycin-induced fibrosis and experimental chronic graft-versus-host disease.¹ These preclinical data suggest that riociguat has vascular and antifibrotic properties that may be beneficial in dcSSc, potentially delaying disease progression.

Objectives: RISE-SSc (NCT02283762) is an international, multicentre, randomised, double-blind, placebo-controlled, parallel-group, Phase II study to assess the efficacy and safety of treatment with riociguat versus placebo in patients with dcSSc. The primary efficacy endpoint is change in modified Rodnan skin score (mRSS) from baseline to Week 52.

Methods: Eligibility criteria include a disease duration of ≤ 18 months and mRSS of 10–22, parameters identified to be predictors of progression of skin fibrosis.² Patients were randomised (1:1) to treatment with riociguat (adjusted up to 2.5 mg tid over the first 10 weeks) or placebo for a total of 52 weeks. The double-blind phase will be followed by an open-label extension phase for up to 6 years where all patients receive riociguat.

Results: Study recruitment was completed on 30 Dec 2016 and 121 patients have been randomised. Mean $\pm$ SD age at inclusion was 50.7 $\pm$ 12.3 years (range 18–79) and 76% (n=92) were female. At baseline the mean $\pm$ SD mRSS was 16.8 $\pm$ 3.7 (range 10–22) and disease duration was 8.8 $\pm$ 5.5 months (range 0.5–18.5). Almost 70% of the patients tested positive for either anti-SCL-70 (43%) or anti-RNA polymerase III (25%) antibodies, while 32% of patients tested negative for these two antibodies. Swollen and tender joint counts were present in 38 (31.4%) and 51 (42.1%) patients, respectively, while 35 patients (28.9%) had tendon friction rubs. Digital ulcers were present in 15 patients (12.4%) at baseline.

Conclusions: RISE-SSc has a unique trial design with a cohort enriched for disease progressors,² which is reflected in the baseline characteristics of the randomised patients. Results of the RISE-SSc study will provide data to inform not only the efficacy, safety and tolerability of riociguat treatment in patients with dcSSc, but also the feasibility of a new trial design based on cohort enrichment.

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Disclosure of Interest: D. Khanna Grant/research support from: Bayer, BMS, Genentech/Roche, Sanofi-Aventis, NIH K24AR063120, Consultant for: Actelion, Bayer, Covis, Cytosil, EMD Serono, Genentech/Roche, Gilead, GSK, Sanofi-Aventis, Y. Allanore Grant/research support from: BMS, Genentech-Roche, Inventiva, Pfizer, Sanofi, Consultant for: Actelion, Bayer, Biogen, Genentech-Roche, Galapagos, Medac, Pfizer, Sanofi, Servier, UCB, C. Denton Grant/research support from: CSL Behring, Bayer, GSK, Inventiva, Consultant for: Actelion, Bayer, GSK, Merck-Serono, Genentech-Roche, Inventiva, Sanofi-Aventis, Boehringer Ingelheim, M. Matucci-Cerinic Grant/research support from: BMS, Pfizer, Actelion, MSD, Lilly, Speakers bureau: Pfizer, BMS, Actelion, Lilly, J. Pope Grant/research support from: Bayer, Merck, Consultant for: BMS, Roche, Bayer, Merck, Z. Yao Employee of: Bayer Healthcare Company Ltd, Beijing, China, J. Curram Shareholder of: Bayer AG, Employee of: Bayer plc, J. de Oliveira Pena Employee of: Bayer US LLC, O. Distler Grant/research support from: Actelion, Bayer, Boehringer Ingelheim, Pfizer, Sanofi, Consultant for: 4 D Science, Actelion, Active Biotech, Bayer, Biogen/Dec, BMS, Boehringer Ingelheim, ChemomAb, EpiPharm, espeRare foundation, Genentech/Roche, GSK, Inventiva, Lilly, medac, Mepha, MedImmune, Mitsubishi Tanabe Pharma, Pharmacoclics, Pfizer, Sanofi, Serodapharm, Sinox, Speakers bureau: AbbVie, iQone Healthcare, Mepha