Methods: We enrolled SSc patients without PH from January 2016 to December 2019 in our institution. SSc patients were diagnosed according to EULAR classification criteria in 2013. ILD was detected by chest CT scans. We assessed severity of ILD with pulmonary function tests (PFT). Abnormal PFT was defined as vital capacity (%VC) or diffusion capacity (DLCO) < 70%. NFC abnormalities were detected with "OptiPiX capillaroscopy Clinic 1.7.x" and the number of capillaries was measured per 1mm in 2nd to 5th fingers of both hand. We defined enlarged and giant capillaries as >30 µm and >50 µm, respectively.

Results: We enrolled 59 SSc patients (54 females, 5 males). Mean age is 65.0 ± 8.0 years. Thirty-one patients (52.5%) were complicated with ILD. Mean capillary counts are 6.6/mm. The number of patients with each NFC abnormalities (enlarged capillaries, giant capillaries, microhemorrhages, ramified, avascular areas) are 42, 32, 48, 38, and 33 cases, respectively. Two cases did not have NFC abnormalities. SSc patients with giant capillaries had fewer ILD complications (p < 0.05, odds ratio 0.183 [0.059 – 0.57]). Other NFC abnormalities were not associated with ILD in SSc patients. We inspected %VC of 23 patients and DLCO of 20 patients with ILD. Eleven patients had abnormal PFT (5 patients had abnormal %VC and 9 patients had abnormal DLCO). Most of them had not enlarged capillaries than patient with normal PFT (odds ratio 0.11 [0.016 – 0.81]). Other NFC abnormalities including giant capillaries were not associated with abnormal PFT.

Conclusion: We investigated the relationship between NFC abnormalities and ILD complications in SSc patients. NFC abnormalities are associated with ILD complication and severity of ILD. It was suggested that no giant capillary in SSc patients may predict ILD complication. Moreover, no enlarged capillary may predict the severe ILD.

References:

- [1] Valeria Riccieri et al. Systemic sclerosis patients with and without pulmonary arterial hypertension: a nailfold capillaroscopy study. *Rheumatology*, Volume 52, Issue 8, 1 August 2013, Pages 1525–1528
- [2] Maurizio Cutolo et al. Nailfold Videocapillaroscopic Features and Other Clinical Risk Factors for Digital Ulcers in Systemic Sclerosis: A Multicenter, Prospective Cohort Study. *Arthritis Rheumatol.* 2016 Oct; 68(10): 2527–2539.

	With ILD(n=31)	Without ILD(n=28)	P
enlarged capillaries	19	23	0.08
giant capillaries	11	21	0.002
microhemorrhage	25	23	0.88
ramified	21	17	0.57
avascular areas	19	14	0.38

Table1. each NFC abnormalities in SSc patients

	Patients had normal PFT(n=12)	Patients had abnormal PFT(n=11)	P
enlarged capillaries	10	4	0.02
giant capillaries	5	3	0.47
microhemorrhage	9	9	0.69
ramified	9	8	0.90
avascular areas	10	6	0.13

Table2. each NFC abnormalities in SSc patients with ILD

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.5623

AB0593 DOES REALLY EXIST MIXED CONNECTIVE TISSUE DISEASE?

L. Montolio-Chiva¹, J. Narváez², M. Pascual², H. S. Park³, A. V. Orenes Vera¹, E. Flores¹, J. J. Alegre-Sancho¹, I. Castellví³, J. M. Nolla². ¹Universitary Peset Doctor Hospital, Valencia, Spain; ²Universitary Bellvitge Hospital, Barcelona, Spain; ³San Pau Hospital, Barcelona, Spain

Background: Currently, most authors accept that mixed connective tissue disease (MCTD) is an independent entity, although there are those who argue that it is actually an overlap syndrome or an undifferentiated early phase of another systemic autoimmune disease (SAD).

Objectives: To analyze the long term evolution of a serie of patients with MCTD.

Methods: Observational, retrospective and multicenter study in patients with MCTD (diagnostic criteria of Alarcón-Segovia et al), followed for a minimum of 2 years.

Results: Fifty-five patients (49 women) with a median age at diagnosis of 38±14 years and with a follow up time (median) of 101 months (range, 24-237 months with a total of 501.2 patients-year) were identified.

At the end of the follow-up period, only 27% (15/55) of the patients kept on fulfilling MCTD criteria. In the remaining 73% (40), 40% (22) had been differentiated to systemic lupus erythematosus (SLE), 13% (7) to systemic sclerosis (SSc) and 20% (11) developed an overlap syndrome [SSc+SLE in 8 cases and SSc+rheumatoid arthritis (AR) in 3]. In 8% of these patients, a secondary Sjögren's syndrome was diagnosed during the follow-up period. The average score in patients who met the EULAR/ACR 2013 criteria for SSc was 11 (minimum 9 - maximum 16) and the average time elapsed from the diagnosis of MCTD to meet SSc criteria was 64.4 months (interquartile range [IQR] 25-75%: 10-127 months).

Applying the 2012 SLICC criteria, only 24 patients of those initially diagnosed as MCTD ended up meeting SLE criteria. The average score in these patients was 5.6 (4-9) and the average time elapsed from the diagnosis of MCTD until fulfilling the SLICC criteria was 39 months (IQR 25-75%: 6-28). When we apply the new ACR/EULAR 2019 criteria, the percentage of patients who meet SLE criteria increased to 30%, with an average score of 17.3 (10-38). The average time elapsed since the diagnosis of MCTD until meeting the new SLE criteria was reduced to 17 months (IQR 25-75: 0-10).

In the multivariate study, the presence of sclerodactyly (OR: 2.91; IC 95% 1.90 - 4.1, p= 0.001) and esophageal involvement (OR: 2.05; IC 95% 1.14–3.66, p=0.016) were associated with the evolution to SSc. Any predictor of evolution to SLE was identified.

Conclusion: Only slightly more than a quarter of patients initially diagnosed as MCTD maintain this diagnosis during the follow-up. The majority, ended up evolving towards to another SAD, fundamentally SLE and SSc. The new ACR/EULAR 2019 criteria seems to be more sensitive than the SLICC 2012 criteria for diagnose SLE in these patients.

Disclosure of Interests: L. Montolio-Chiva: None declared, J. Narváez: None declared, Maria Pascual: None declared, Hye Sang Park: None declared, Ana V Orenes Vera: None declared, Eduardo Flores: None declared, Juanjo J Alegre-Sancho Consultant of: UCB, Roche, Sanofi, Boehringer, Celltrion, Paid instructor for: GSK, Speakers bureau: MSD, GSK, Lilly, Sanofi, Roche, UCB, Actelion, Pfizer, Abbvie, Novartis, Iván Castellví: None declared, Joan Miquel Nolla: None declared

DOI: 10.1136/annrheumdis-2020-eular.3843

AB0594 EFFECTIVENESS OF RITUXIMAB IN CSDMARDS-RESISTANT ACTIVE MIXED CONNECTIVE TISSUE DISEASE

L. Montolio-Chiva¹, J. Narváez², J. J. Alegre-Sancho¹, J. Lluch Pons², A. V. Orenes Vera¹, I. Vázquez-Gómez¹, M. Mora², X. González², C. Marco², J. Rodríguez², M. Romero², J. M. Nolla². ¹Universitary Peset Doctor Hospital, Valencia, Spain; ²Universitary Bellvitge Hospital, Barcelona, Spain

Background:

Objectives: To evaluate rituximab (RTX) effectiveness and safety in treating patients with refractory mixed connective tissue disease (MCTD).

Methods: Open observational study including patients with refractory MCTD (active disease despite treatment with glucocorticoids and csDMARDs) from two third-level hospitals who had been treated with RTX (off-label use) from January 2001 to December 2019.

Results: Thirteen patients (all women) were included, with a mean age of 32 years (SD: 10, range 17-50) and a median time of evolution of the disease of 55 months (SD: 34.3; range 5-98 months). The main indication for initiating treatment with RTX was refractory arthritis (100%), most of the times accompanied by other features of the disease including shrinking lung syndrome (2), fibrosing progressive non-specific interstitial pneumonia (FP-NSIP) (1), recurrent serositis (2), glomerulonephritis (GMN) (2), lymphadenitis (1) and immune thrombocytopenic purpura (ITP) (1). All patients were treated with RTX at rheumatoid arthritis dosage while the baseline immunosuppressive treatment (methotrexate, azathioprine, mycophenolate, leflunomide or tacrolimus) remained unchanged. Hydroxychloroquine was also associated in 8 of the patients. The follow-up time (median) after starting RTX was 118 months (range, 65-177 months, with a total of 132.6 patient-years of follow-up) and the mean number of cycles of treatment was 4.2 (range, 1-15), with a variable interval (from 6 to 12 months). After the first RTX cycle, a partial or complete response was achieved in 92% of the patients. A significant improvement in the mean DAS28-ESR was observed (initial: 4.56 ± 1.6 / final: 2.21 ± 0.85; p=0.008). In all but one patient, who had previously failed to 2 anti-TNFα