Methods: Observational study using data from the Portuguese Rheumatic Diseases Register (Reuma.pt/Myositis protocol). Data extracted included demographic variables, clinical features and immunological expression of the disease.

Results: 17 patients were identified. All met the criteria for AS according to Connors criteria, while 3 did not met according to the Solomon criteria. Mean age at diagnosis was 60.1 years (26 - 80) and 76.5% were female. Mean follow-up time was 2.8 (0.5-9) years. Only 3 patients had history of smoking in the past. The autoantibodies expressed were anti-Jo1 (n=12), anti-PL12 (n=2), anti-OJ (n=2) and anti-PL7 (n=1). 4 patients positive for anti-Jo-1 also expressed anti-Ro52. The clinical information and treatment are described in table 1. One patient presented as a paraneoplastic syndrome associated with anti-Jo-1.

Table 1. Demographic and clinic characteristics of our cohort. AZA – azathioprine; CYC – Cyclophosphamide; DM – dermatomyositis; GS – Gottron's sign; ILD – Interstitial lung disease; PM – Polymyositis; NSIP - Nonspecific interstitial pneumonia; MH – mechanic hands; MMF – Mycophenolate mofetil; MTX – Methotrexate; PDN – prednisolone; RP – Raynaud phenomenon; RTX – Rituximab; UIP - Usual interstitial pneumonia; Y – yes; N – no. Connors criteria – anti-ARS plus one or more of RP, MH, arthritis, ILD, fever. Solomon criteria – anti-ARS plus 2 major criteria or 1 major and 2 minor criteria. *induction therapy.

Case	Serology	Major criteria (ILD) [NSIP/UIP/ non-specific] or PM/ DM)	Minor criteria (Arthritis, MH, RP)	Other manifestations	Connors (2010)/ Solomon (2011) criteria	PDN	DMARDcs (MTX/MMF/ AZA)	CYC/ RTX
1	Jo1	PM	MH, arthritis	GS	Y/Y	Y	MMF	-
2	Jo1	NSIP	RP, MH	Asthenia	Y/Y	Y	AZA	-
3	Jo1	NSIP	MH, arthritis	Calcinosis, asthenia	Y/Y	Y	AZA	-
4	Jo1	NSIP; DM	-	GS	Y/Y	Y	-	-
5	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MTX	-
6	Jo1	UIP	RP, arthritis	Asthenia	Y/Y	Y	-	RTX
7	Jo1/Ro52	NSIP; PM	arthritis	-	Y/Y	Y	MMF	CYC*
8	Jo1	NSIP; PM	-	-	Y/Y	Y	MMF	-
9	Jo1/Ro52	NSIP; PM	-	-	Y/Y	Y	AZA	-
10	PL7	-	RP, arthritis, MH	Dysphagia	Y/N	Y	-	-
11	Jo1	-	RP	Asthenia	Y/N	-	-	-
12	Jo1/Ro52	PM	MH, arthritis	GS, asthenia	Y/Y	Y	MMF	-
13	PL12	PM	RP, arthritis	GS, asthenia	Y/Y	Y	MMF	-
14	Jo1	UIP	RP, arthritis	Weight loss, asthenia	Y/Y	Y	-	RTX
15	OJ	Non-specific pattern	-	-	Y/N	Y	MMF	-
16	OJ	NSIP	-	-	Y/Y	Y	-	-
17	PL12	PM	RP, MH	-	Y/Y	-	-	-

Conclusion: The most frequent autoantibody was anti-Jo-1, which is consistent with the literature. Interestingly, patients with anti-PL, usually described as having severe lung disease, in our series do not have it. Additionally, we found a trend for a younger age at diagnosis in Jo1 positive patients and remarkably more than half of these patients have been diagnosed with ILD, being the NSIP pattern the most frequently reported.

Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.4857

AB0591 ANALYSIS OF A COHORT OF PATIENTS WITH SYSTEMIC SCLEROSIS AND INTERSTITIAL LUNG DISEASE

L. Mendez Diaz¹, R. J. Gil Velez¹, I. Madroñal García¹. ¹Seville, Seville, Spain

Background: Interstitial lung disease (ILD) is a common manifestation of systemic sclerosis that is associated with early mortality. The development or progression of ILD can occur at any time, so patients should be monitored regularly, particularly in the first years after diagnosis.

Treatment should be considered when the disease is clinically significant, particularly when there is evidence of progression based on a decrease in lung function, progression of fibrosis on the HRCT or worsening of respiratory symptoms.

Objectives: -To relate the type of systemic sclerosis (SS) with pulmonary involvement with the radiological pattern.

-To study if there is a relationship between the antibodies and the aforementioned affection.

Methods: Retrospective descriptive study of patients treated in our Hospital (2009-2019) by the Rheumatology and Internal Medicine department diagnosed with systemic sclerosis and interstitial lung disease.

The data were obtained through the review of medical records.

We have included data from patients who have a diagnosis of limited or diffuse systemic sclerosis or overlap with interstitial lung involvement.

Results: Of the 213 patients with systemic sclerosis in our database 43 had interstitial lung involvement (20.2%). 79% of the patients with ILD (34) had a non-specific interstitial pneumonia type (NSIP) radiological pattern and 21% of the patients (9) had a pattern of usual interstitial pneumonia (UIP)

Among the patients with ILD with a NSIP -type radiological pattern, 19 patients were diagnosed with diffuse SS, 9 patients with overlap syndrome and 6 with limited SS. Of the patients with ILD with radiological pattern type UIP, 5 patients were diagnosed with diffuse SS, 3 patients with overlap syndrome and 1 patient was diagnosed with limited SS.

TABLE 1. RELATIONSHIP BETWEEN SS TYPE AND RADIOLOGICAL PATTERN

	NSIP	UIP
LIMITED SS	6 (17.6%)	1 (11.1%)
DIFFUSE SS	19 (55.8%)	5 (55.5%)
OVERLAP	9 (26.4%)	3 (33.3%)

Among the patients with the NSIP pattern, 17 had positive SCL70 antibody, 3 positive ANA patients and 1 patient had positive anti-centromere antibody.

Of the patients with UIP type interstitial pneumopathy, 8 patients had anti-SCL70 antibody, 3 patients ANA positive antibody and 2 patients anti-centromere positive antibody.

TABLE 2. RELATIONSHIP BETWEEN TYPE OF AB AND RADIOLOGICAL PATTERN

	NSIP	UIP
Anti SCL70	17	8
Anti centromere	1	2
ANA	3	3

Regarding treatment, 21 patients were taking Mycophenolate, 16 patients required cyclophosphamide and 6 patients rituximab.

No patient in our cohort died due to interstitial lung disease.

Conclusion: The data obtained are consistent with what is collected in the medical literature.

The subtype of scleroderma most related to ILD was diffuse SS. The most frequent antibody was anti-SCL 70.

Regarding the treatment, the most used in ILD in our center was the mycophenolate.

From our sample analyzed when applying the likelihood ratio (RV) a value of 47,186 is obtained, which has an associated probability of 0, which is less than 0.05, leads to reject the null hypothesis (there is no dependence between antibodies and type of radiological pattern of ILD in SS), concluding that there is dependence between the analyzed variables.

After this analysis, we can conclude that in our sample there is a relationship between the type of interstitial pneumopathy pattern and the antibody present in patients with SS.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.2673

AB0592 NAILFOLD CAPILLARY ABNORMALITIES PREDICT INTERSTITIAL LUNG DISEASE (ILD) COMPLICATION IN SYSTEMIC SCLEROSIS PATIENTS

T. Miyagi¹, T. Kameda¹, S. Nakashima¹, H. Shimada¹, R. Wakiya¹, M. Kato¹, M. Mahmoud Fahmy Mansour¹, K. Sugihara¹, Y. Ushio¹, H. Dobashi¹. ¹Kagawa University, Division of Hematology, Rheumatology and Respiratory Medicine, Internal Medicine, Kagawa, Japan

Background: Systemic sclerosis (SSc) have various organ involvements including pulmonary hypertension (PH), digital ulcers (DU), and interstitial lung disease (ILD). On the other hand, Nailfold capillary (NFC) abnormalities (enlarged/giant capillaries, fresh or old hemorrhages, avascular areas, ramified/bushy capillaries) detected by capillaroscopy are included in ACR/EULAR classification criteria for SSc as one of important findings. In addition, many studies have reported the relationship between NFC abnormalities and organ involvements (DU, PH) [1][2]. However, there are a few reports about the relationship between NFC abnormalities and ILD.

Objectives: We clarify the association with NFC abnormalities and ILD in SSc patients.

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