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

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AB0593

DOES REALLY EXIST MIXED CONNECTIVE TISSUE DISEASE?

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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.

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AB0594

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

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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

DAS28-ESR clinical remission or low activity was achieved, generally from week 16 to 20, although relapses were frequent and all cases need retreatment after 6-9 months. In 4 patients, RTX retreatment dosage was optimized to 1 g/cycle. The 3 patients with pulmonary involvement showed stabilization (2 cases) or improvement (1) of the lung function (*as defined by the American Thoracic Society*). In patients with GMN, renal response to RTX treatment was complete in a patient and partial in the other. The patient with ITP entered remission after the first RTX cycle and no more cycles were needed. Response in patients with serositis and lymphadenitis was also complete and maintained. Moreover, the glucocorticoid doses were reduced to less than half of the initial dose in all cases. At the end of the follow-up, 7 out of the 13 patients (54%) were still being treated with RTX. For the remaining 6 patients, RTX was withdrawn because of primary failure (1), recurrent bacterial infections (2), gestational desire (2) sustained remission (1).

Conclusion: According to our preliminary results, RTX seems to be effective and relatively safe in patients with csDMARDs-resistant active MCTD.

Disclosure of Interests: L Montolio-Chiva: None declared, J. Narváez: 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, J Lluch Pons: None declared, Ana V Orenes Vera: None declared, I Vázquez-Gómez: None declared, Maribel Mora: None declared, Xavier González: None declared, Carla Marco: None declared, Jesús Rodríguez: None declared, Montserrat Romera: None declared, Joan Miquel Nolla: None declared

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AB0595

EFFECTIVENESS OF TOPICAL SODIUM TIOSULFATE FOR THE TREATMENT OF CALCINOSIS-ASSOCIATED CUTANEOUS ULCERS IN PATIENTS WITH SYSTEMIC SCLEROSIS

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Background: Treatment of calcinosis associated with systemic sclerosis (SSc) mainly involves the use of systemic therapies, which often have limited efficacy. However, little attention has been paid to local treatment, which is especially useful when associated with skin ulcers.

Objectives: To show our experience with topical sodium thiosulfate (TST) for the treatment of calcinosis-associated cutaneous ulcers in patients with SSc.

Methods: Descriptive analysis of a case series of patients with SSc and calcinosis-associated skin ulcers treated with TST. Wound management procedure: wounds and perilesional skin cleaning and disinfection is performed and, if needed, additional debridement. TST is compounded at 25% as w/o emulsion, for extensive calcinosis, or as beeler-base or cold-cream ointment, for limited calcinosis. Wounds are then covered with a polymeric foam dressing. This cure in moist healing environment shows some advantages over the dry cure (exudate control without damaging the perilesional skin, protection against contamination, and reduction of the needed cures, healing time and pain).

Results: Nine patients (7 women) with calcinosis-associated skin ulcers and SSc were included: 2 patients with diffuse SSc (DcSSc), 6 with limited SSc (LcSSc) and 1 with overlap syndrome. Median age was 60 years (IQR 20). 6 patients had localized wounds and 3 had extensive involvement and/or tumoral calcinosis which had been refractory to systemic treatment with diltiazem, colchicine, zoledronate, rituximab, and/or acenocoumarol and had suffered recurrent superinfections. Follow-up results of more than 3 months are available for 8 patients, who have been on TST a median time of 9 months (IQR 8.25). They have shown clinical improvement (disappearing of many calcinosis foci and partial or complete healing of the ulcers together with an improvement in pain, function, quality of life and satisfaction of the patients). Radiological improvement was also observed in 1 case. No TST related adverse effect has been detected, except for slight maceration of the wound edges due to the ointment preparation, which was resolved by protecting these with zinc oxide cream

Conclusion: In our experience, treatment with TST for calcinosis-associated skin ulcers in patients with SSc is an effective, safe and easily implementable therapeutic alternative in clinical practice.

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MSD, Novartis, Janssen, Bristol Myers Squibb, UCB Pharma, D Ybáñez-García Speakers bureau: Lilly, Roche, Sanofi, 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

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AB0596

PREDICTORS, LONG TERM CLINICAL AND TREATMENT OUTCOMES IN SOUTH ASIAN PATIENTS WITH IDIOPATHIC INFLAMMATORY MYOSITIS: A SINGLE CENTER STUDY

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Background: Idiopathic inflammatory myositis (IIM) are a heterogeneous group of immune-mediated disorders with varied presentations and multiple organ involvement. Data on long term outcome among South Asian patients with IIM is sparse.

Objectives: To study the long term clinical outcome, treatment responses and factors predicting outcome among adult patients with IIM

Methods: Patients diagnosed as 'Idiopathic Inflammatory Myositis' under the department of Clinical Immunology and Rheumatology at CMC, Vellore, India were screened retrospectively. Patients aged 18 years and above, satisfying Bohan and Peter criteria, having follow up of one year or more with atleast two outpatient or inpatient visits between January 2010 and April 2019 were included in this study. Those patients with connective tissue disease associated myositis were not included. Details on muscle weakness, extramuscular involvement, muscle enzymes and treatment administered were recorded at baseline, 3, 6, 12, 18, 24 months and yearly thereafter. After assessing their cumulative response, categorization of patients into complete and partial responders was done. Complete responders were defined as patients with persistent muscle power of more than 4/5 and/or MMT 8 more than 76/80, complete resolution of skin, articular and lung involvement (if any) as well as muscle enzymes less than twice the upper limit of normal without any documented flares during the entire follow up period. Patients not satisfying the said criterias were grouped as Partial responders. Disease free survival duration was also analyzed.

Results: Out of 310 patients of IIM identified, 187 (60.3%) patients satisfied the inclusion criteria. Women were 2.2 times more than men and mean age at symptom onset was 35.7±12.6 years. Dermatomyositis was the predominant myositis subtype seen. All patients were put on steroids with the mean dose being 45.9 ± 18.6 mg/day. At baseline, the key immunosuppressants used were methotrexate in 44.9% and mycophenolate in 37.6% patients. The median follow up duration was 48 (25-80) months. An associated malignancy was diagnosed in 3.2% after a median duration of 24.5 months. Five patients expired after a median duration of 80 months from diagnosis. Normal muscle power was attained in 76.1% patients and 88.6% were vocational by the last follow up visit. Steroids were discontinued in 56.7% patients after a median duration of 24 months (p=0.0002). Discontinuation of the immunosuppressant was feasible in 10.2% patients after a median duration of 44 months. Assessment of

Figure 1. Disease free survival plot of patients with IIM