sets of 93 pairs of TCZ/controls were generated. Comparison between groups showed consistent effects of TCZ across all pre-defined primary and secondary endpoints: mRSS was lower in the TCZ group (mean difference (95% confidence interval (CI)) -1.8 (-4.79 to 1.19), $p=0.24$, Figure 1A). Similarly, FVC % predicted was higher in the TCZ group mean difference (2.25, 95% CI -4.57 to 9.06), $p=0.51$, Figure 1B). Considering secondary endpoints, the percentage of skin progressors as well as lung progressors at follow up was lower in the TCZ group (odds ratio OR 0.67 (95% CI 0.07 to 6.41), $p=0.74$ and OR 0.53 (95% CI 0.16 to 1.7); $p=0.2$, respectively. Consistently, the percentage of regressors for skin (OR 1.6 (95% CI 0.56 to 4.54), $p=0.38$) and for lung (OR 1.74 (95% CI 0.66 to 4.58), $p=0.26$) was higher in TCZ. These results were robust regarding the sensitivity analyses. Safety analysis confirmed previously reported adverse event profiles.

Conclusion: In this large, observational, controlled, real-life EUSTAR study, effectiveness of TCZ did not reach statistical significance compared to standard of care treatment but showed consistent positive effects of TCZ on skin and lung fibrosis across all pre-defined primary and secondary endpoints confirming data from recent RCTs.

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Figure 1. mRSS and FVC at follow up (123 months)

- A) Mean mRSS (95% CI) TCZ 10.68 (8.96 to 12.4) vs control 12.5 (10.5 to 14.5), between group mean difference -1.8 (-4.79 to 1.19), $p=0.24$
- B) Mean FVC, % predicted (95% CI) TCZ 89.22 (84.77 to 93.67) vs control 86.97 (82.43 to 91.52), between group difference 2.25 (-4.57 to 9.06), $p=0.51$

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Topadur, Target Bioscience and UCB. Patent issued "mir-29 for the treatment of systemic sclerosis" (US8247389, EP2331143)., Consultant of: Oliver Distler has/had consultancy relationship and/or has received research funding in the area of potential treatments for systemic sclerosis and its complications from (last three years): Abbvie, Acceleron Pharma, Amgen, AnaMar, Arxx Therapeutics, Baeon Discovery, Blade Therapeutics, Bayer, Boehringer Ingelheim, ChemomAb, Corbus Pharmaceuticals, CSL Behring, Galapagos NV, Glenmark Pharmaceuticals, GSK, Horizon (Curzion) Pharmaceuticals, Inventiva, iQvia, Italfarmaco, iQone, Kymera Therapeutics, Lilly, Medac, Medscape, Mitsubishi Tanabe Pharma, MSD, Novartis, Pfizer, Roche, Sanofi, Serodapharm, Topadur, Target Bioscience and UCB. Patent issued "mir-29 for the treatment of systemic sclerosis" (US8247389, EP2331143)., Grant/research support from: The study was partially supported by a grant from Roche. Roche was not involved in analysis or interpretation of the results.

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POS0862 NAILFOLD CAPILLAROSCOPY IN UNDIFFERENTIATED AND MIXED CONNECTIVE TISSUE DISEASES

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Background: Microvascular damage is a frequent feature in connective tissue diseases (CTDs) and can be easily detected through nailfold videocapillaroscopy (NVC) (1,2). Mixed and Undifferentiated connective tissue diseases (MCTD and UCTD) do not show a specific and unique NVC pattern (3). However, a variety of microvascular abnormalities can occur in these two CTDs, both non-specific or specific for the scleroderma like-pattern (3-5).

Objectives: To retrospectively assess and compare nailfold microangiopathy observed by NVC in MCTD and stable UCTD versus primary Raynaud's phenomenon (PRP) (6). In addition, the aim was to correlate NVC findings with serum levels of autoantibodies (Abs) against extractable nuclear antigen (ENA) detected in UCTD.

Methods: Files of forty-six MCTD patients (Kasukawa's criteria) (mean age 42.8±16 SD years), forty-seven UCTD patients (mean age 47.7±16.1 SD years), fifty-one PRP (mean age 45.9±17.3 SD years) were retrospectively evaluated in the study. Among UCTD and MCTD patients 95% of both showed Raynaud's phenomenon. Main NVC parameters (i.e. dilated capillaries, giant capillaries, microhemorrhages, abnormal shapes and number of capillaries) and related semiquantitative scale (score 0–3 for every parameter), were analyzed and compared between the two distinct CTD groups and PRP. Furthermore, ENA Abs (in particular, Ro/SSA, La/SSB, Scl70 and Jo1) were evaluated. The CTD patients were receiving different immunosuppressive treatments. Statistical analysis was performed by non-parametric tests.

Results: Among UCTD group, 36% of patients showed a normal NVC pattern, 53% had non-specific NVC abnormalities and 11% had a scleroderma like-pattern. The latter was significantly more frequent in MCTD than in UCTD ($p<0.001$), in fact 22 out of 46 (48%) MCTD patients presented a scleroderma-like pattern. On the other hand, normal pattern or non-specific NVC abnormalities were respectively found in 9% and 43% MCTD patients. Therefore, CTD patients showing giant capillaries, abnormal shapes (i.e. angiogenesis) and lower capillary density were significantly more affected by MCTD than UCTD ($p<0.001$). Finally, the absolute number of capillaries was found significantly lower in MCTD versus UCTD patients (mean 7±1.7 SD vs mean 9.2±1 SD, respectively, $p<0.001$). Not any statistical correlation was observed between NVC parameters and specific Abs ENA in UCTD. PRP showed a normal NVC pattern in 2% and non-specific capillary abnormalities in 98%, (including dilated capillaries and microhemorrhages).

Conclusion: NVC features in UCTD patients seem very close to the pattern observed in PRP (mostly non-specific capillary abnormalities), conversely in MCTD the scleroderma-like pattern was found significantly prevalent together with a significant capillary number reduction. The transition from the scleroderma-like to the scleroderma pattern (mean systemic sclerosis) is matter of actual investigation.

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