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FRI0412 THE POSITIVE EFFECT OF RITUXIMAB IN PULMONARY FIBROSIS OF SYSTEMIC SCLEROSIS

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Background: Pulmonary fibrosis is one of the gravest manifestations of Systemic Sclerosis (SSc) and conventional DMARDs therapy has not shown any particular positive effect.
Objectives: Our goal was to see whether the elimination pf B - lymphocytes through use of anti – CD20 Mab, Rituximab (RTX) would offer to the improvement of the pulmonary function of SSc patients.
Methods: We studied 23 SSc patients with pulmonary fibrosis, who received treatment with RTX (n=12) or DMARDs treatment (n=11) for 1 to 3 years (1,9 years). Conventional therapy included azathioprine (n=4), mycophenalate (n=6) and methotrexate (n=2). RTX – treated patients were recorded with FVC improvement in the first year of treatment (FVC 81,3 +/- 12,6 vs FVC 87,4 +/- 11,3 out on the onset of the study and at the first year respectively, p=0,02) when on the other hand DMARDs treated patients didn't show any FVC improvement at all.
Results: All RTX – treated patients did not present any lung HRCT deterioration imaging, in contrast to DMARDs – treated patients who were also submitted to lung HRCT each year of the study, all showing signs of CT imaging deterioration. Similar findings, as far as FVC was concerned, were recorded at the 3rd year of the study (RTX patients, n=6 and DMARDs patients, n=11). 3rd year RTX FVC was 92,6 +/-13,2 vs 80,7 +/-11,8 at the onset of the study, p=0,04 when DMARDs treated patients were all presented with worst 3rd year FVC compared to their primary FVC, p<0,01.
Conclusions: Our small cohort of SSc patients with pulmonary fibrosis shows that it is possible that rituximab may be proven helpful to at least prevent the deterioration of the interstitial pulmonary fibrosis of Systemic Sclerosis. The pathophysiology of this particular fibrosis and the possible molecular role of B-lymphocytes in the inversion of this procedure need yet to be furtherly explored.
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FRI0413 ACUTE EFFECT OF ILOPROST ON PERIPHERAL CIRCULATION AS ASSESSED BY VIDEOCAPILLAROSCOPY AND 22-MHZ POWER DOPPLER ULTRASONOGRAPHY

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Background: Vascular involvement is a hallmark of systemic sclerosis (SSc) and it is responsible for some of the most common complications of the disease such as Raynaud's phenomenon (Rp), digital ulcers (DUs) and pulmonary arterial hypertension. I.V. iloprost (ILO), a prostacyclin analogue, has been shown to be effective in reducing Rp severity, DUs healing and preventing [1].
Objectives: To assess the acute effect of ILO on acral circulation as assessed by nailfold videocapillaroscopy (NVC) and 22-MHz Power Doppler ultrasound (PDUS).
Methods: 44 SSc consecutive patients fulfilling the 2013 EULAR classification criteria were enrolled. Each patient was evaluated before and immediately after I.V. ILO administration (0.5–2.0 ng/Kg/min for 6 consecutive hours). PDUS was performed at the 3rd and 4th finger of the dominant hand after exclusion of ulnar artery occlusion (UAO). In case of UAO non-dominant hand was examined. Ultrasound investigation was performed with Esaote MyLab 70 XVG by means of linear array transducer (10–22 MHz). Power Doppler settings were standardized (Doppler frequency 14.3 MHz, Gain 55%, PRF 750 Hz). PDUS measurements included sagittal scan of nailbed and fingertip qualitatively graded from 1 (no signal) to 4 (marked hyperemia) [2], and resistivity index (RI) of ulnar and radial proper digital arteries. Capillary width (sum of capillary width/mm) was calculated by NVC with magnification 200X performed on two images of the same digits examined by PDUS.
Results: The study population included 44 SSc patients, 40 (90.9%) women, 35 (79.5%) limited cutaneous SSc, median age 60.2 years old and median disease duration 8 years. 19 (43.2%) had history of DUs, among them 15 had experienced more than one DUs and 1 had active DU at the moment of evaluation.
Semiquantitative perfusion score of sagittal scan of nailbed and fingertip pre- and post-therapy are shown in Table 1.
Changes in RI and capillary width pre- and post-infusion are reported in Table 2.
Conclusions: A statistically significant post-infusion rise in RI, fingertip and nailbed PDUS grade were found. Capillary width was also increased, but it was not statistically significant. As such, these novel results indicate that ILO, alongside its clinical effect, is able to enhance vascularization even at the most peripheral levels.

Table 1

	Pre-infusion		Post-infusion		OR (95% CI)
	%	Cumulate %	%	Cumulate %	
Fingertip PDUS					4.21 (2.12–8.35)
Grade 1	29.55	29.55	14.77	14.77	p-value<0.001
Grade 2	22.73	52.28	15.91	30.68	
Grade 3	12.50	64.78	12.50	43.18	
Grade 4	35.23	100	56.82	100	
Nailbed PDUS					8.96 (4.15–19.32)
Grade 1	17.05	17.05	6.82	6.82	p-value<0.001
Grade 2	34.09	51.14	13.64	20.46	
Grade 3	18.18	69.32	14.77	35.23	
Grade 4	30.68	100	64.77	100	

Table 2

	Pre-infusion	Post-infusion	Mean difference (95% CI)
Resistivity index	0.773	0.794	0.021 (0.005, 0.037) p-value=0.0122
Capillary width	247.5	257.9	12.321 (-2.528, 27.171) p-value=0.1077

References:
[1] Pope J et al. Cochrane Database Syst Rev. 2000;(2):CD000953.
[2] Newman JS et al. Radiology. 1996,198:582–584.
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FRI0414 CAN ECHOCARDIOGRAPHY REPLACE FOLLOW-UP CARDIAC CATHETERIZATION IN RE-EVALUATION OF PULMONARY ARTERIAL HYPERTENSION? A LONGITUDINAL SINGLE-CENTER STUDY OF 30 CONNECTIVE TISSUE DISEASE PATIENTS

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Background: Transthoracic echocardiography (TTE) is well validated for initial assessment of connective tissue disease patients with suspected pulmonary arterial hypertension (PAH). However, in patients with PAH confirmed by the gold-standard method of right heart catheterization (RHC) the role of TTE in their follow-up is less known.
Objectives: To test the hypothesis that TTE can replace follow-up RHC in connective tissue disease-associated PAH.
Methods: This retrospective study included 30 consecutive patients with systemic sclerosis (n=24) and mixed connective tissue disease (n=6) (mean age±SD: 60±12 years, 87% women), in whom PAH was suggested by TTE and further confirmed by a baseline RHC [pulmonary artery systolic pressure (PASP): 56.8±19.1, range 25–90mmHg; pulmonary vascular resistance (PVR): 5.9±3.8, range 0.7–14.5 Wood units; cardiac output: 4.4±1.3, range 2–7.8 L/min). All 30 patients underwent a second RHC and TTE at follow-up, after 11±6 (range 4–29) months. Ten patients had a 3rd follow-up RHC and TTE 22±7 (range 15–37) months from baseline, thus producing in all 50 pairs of baseline and follow-up measurements. By considering follow-up RHC as the gold-standard, we examined whether clinically meaningful hemodynamic changes (i.e.>15% change from baseline) in either RHC-derived PASP or PVR could be predicted by the corresponding changes from baseline in follow-up TTE. RHC and TTE were always performed, each, by the same examiner.
Results: In 68% of comparisons between baseline and follow-up, the latter TTE measurements could safely replace RHC in terms of PASP estimation. Using Mc Nemars test we confirmed that the two methods did not differ significantly (Table 1). When in addition to changes in PASP, PVR changes were also considered, follow-up TTE could again safely replace the second RHC in 70% patient retests (Table 2). Of note, baseline hemodynamic values or TTE measurements did not differ between patients in whom TTE could replace RHC and those in whom the results of the two methods at follow-up were divergent.

Table 1. McNemars test comparing PASP estimated by repeat TTE to that measured by repeat RHC shows no difference between the two methods

Repeat TTE	Repeat RHC	
	Stable or improved PASP	Deteriorated PASP (>15%)
Stable or improved PASP	25	9
Deteriorated PASP (>15%)	7	9

P=NS (0.8).

Conclusions: In a study where operator-dependent methodological errors are limited we found that the majority of connective tissue disease patients with PAH