Abstract AB0796 – Table 1

Count of FEV1	Column Labels				
Row Labels	31–50	51–70	71–90	91–110	Grand Total
31–50	4	4			8
51–70		25	23	4	52
71–90			14		14
Grand Total	4	29	37	4	74

There was no association between treatment response and age, sex or type of connective tissue disease. Following treatment there was a definite improvement in the clinical aspects of the patient which was assessed by dyspnoea Borg score and cough visual analogue score. Following treatment there was a definite improvement in PFT (FVC, FEV1) as well as 6 min walk test which was also statistically significant. CT pattern of Non specific interstitial pneumonitis also had a better outcome.

Conclusions: Our study proves that there is definite clinical and FVC improvement in CTD-ILD patients after giving treatment with cyclophosphamide. Determinants that can predict treatment outcome are duration of disease, HRCT type, (NSIP), presence of Pulmonary artery hypertension (PAH). Poor treatment response should be anticipated in those with longer duration of disease and UIP type in HRCT, presence of PAH.

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AB0797

A RANDOMISED DOUBLE BLIND PLACEBO CONTROLLED TRIAL TO COMPARE THE EFFICACY OF INITIAL COMBINATION THERAPY VS MONOTHERAPY FOR PULMONARY ARTERIAL HYPERTENSION IN SYSTEMIC SCLEROSIS

P. Dwivedi¹, S.K. Sharma¹, V. Dhir¹, M.K. Rohit², D. Behera³, S. Jain¹. ¹Internal Medicine; ²Cardiology; ³Pulmonary Medicine, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Background: Pulmonary Arterial Hypertension (PAH) is one of the leading cause of death in Systemic Sclerosis (SSc) population. Trials that have studied the efficacy of initial oral combination therapy versus monotherapy for the treatment of PAH in SSc are limited.

Objectives: To study the efficacy and safety of monotherapy (sildenafil) versus initial combination therapy (sildenafil and bosentan) for treatment of SSc related PAH.

Methods: In a single centre, double blind, prospective, randomised, placebo controlled trial, 34 patients of SSc related PAH defined as Pulmonary Artery Systolic Pressures (PASP) >35 mmHg as measured by echocardiography, New York heart Association (NYHA) functional class II and III with forced vital capacity >60% were randomised in 1:1 ratio to sildenafil (20 mg thrice a day) and matched placebo of bosentan to one study arm or to the combination of sildenafil (20 mg thrice a day) and bosentan (62.5 mg twice daily for 4 weeks maximum up to 125 mg twice daily) to the other arm for 24 weeks. The primary end point of the study was to assess the change in pulmonary artery pressures measured by echocardiography at 24 weeks from baseline. The secondary efficacy end points was to compare the change in 6 min Walk Distance (6MWD), Time To Clinical Worsening (TTCW) and adverse events at 24 weeks. Intention to treat analysis was carried for the primary and secondary outcomes. $p < 0.05$ was considered significant. Kaplan Meier survival analysis was done to estimate the time to PAH worsening and hazard ratio was calculated.

Results: The mean change in Pulmonary Artery Systolic Pressures in the monotherapy arm was -1.0 mmHg versus $+2.1$ mmHg in the combination arm ($p = 0.56$). TTCW is prolonged in the combination arm. The mean survival time in the initial combination arm was 23.76 ± 0.59 weeks and 23.28 ± 0.23 weeks in the monotherapy arm. The hazard ratio was 0.73 [95% C.I. 0.04 – 11.7] ($p = 0.87$). The mean change in the 6 min walk distance in the monotherapy arm was 15.88 ± 31.83 m and 25.88 ± 38.25 m in the combination arm ($p = 0.38$). The satisfactory clinical response in the combination arm was 82.3% versus 70% in the

monotherapy arm (O.R = 1.9 [C.I. 0.38 – 9.88] $p = 0.44$). The upfront combination of sildenafil and bosentan was well tolerated with similar rates of adverse events in both group.

Conclusions: In this study we could not demonstrate any significant difference in the efficacy of initial combination therapy of sildenafil and bosentan over sildenafil monotherapy. The patients of systemic sclerosis tolerated the initial combination of sildenafil and bosentan well and the treatment prevented further deterioration. Larger studies with more number of subjects may be required to confirm the results.

Trial Registration- Clinical Trials.gov: NCT03053739

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AB0798

CLINICAL ASSOCIATIONS OF VASCULAR ENDOTHELIAL GROWTH FACTOR AND ITS RECEPTOR 2 TYPE IN SYSTEMIC SCLEROSIS

R. Alekperov^{1,2}, E. Alexandrova², A. Novikov², L. Ananyeva². ¹Moscow Regional Research and Clinical Institute (MONIKI); ²NASANOVA RESEARCH INSTITUTE OF RHEUMATOLOGY, MOSCOW, RUSSIA, Moscow, Russian Federation

Background: Systemic sclerosis (SSc) is characterised by impaired angiogenesis and peripheral microvasculopathy. A major role in the regulation of angiogenesis attributed to vascular endothelial growth factor, which performs its functions through interaction with its receptor 2 type (VEGFR2).

Objectives: The aim of our study was to assess the levels of VEGF in serum and its associations with clinical manifestations in patients with SSc.

Methods: We studied 46 patients with SSc who underwent clinical examination, pulmonary function tests and echocardiography. Serum VEGF-A and VEGFR2 levels were determined by ELISA in 46 SSc patients and 20 healthy controls

Results: Mean VEGF levels were increased in SSc patients compared to controls (212.35 ± 253.93 and 97.74 ± 71.46 pg/ml, respectively; $p = 0.032$). Patients with limited cutaneous SSc ($n = 23$) had higher levels of serum VEGF than those with diffuse cutaneous SSc ($n = 23$) (267.11 ± 268.74 vs 120.40 ± 141.09 pg/ml; $p = 0.012$). Patients with fingertip ulcers ($n = 19$) were found to have higher levels of VEGF than pts without fingertip ulcers ($n = 27$) (214.25 ± 265.93 vs 162.88 ± 188.97) but this difference wasn't significant. Serum VEGF levels were higher in SSc patients with sPAP >30 mmHg than in those with sPAP ≤30 mmHg (286.51 ± 287.42 vs 92.88 ± 108.06 pg/ml; $p = 0.0042$) and correlated with sPAP values ($R = 0.40$; $p = 0.007$). Mean VEGF levels in patients with DLCO <50% ($n = 14$) were significantly higher than those with DLCO ≥50% ($n = 30$): 364.20 ± 381.95 vs 128.55 ± 142.70 pg/ml, respectively ($p = 0.034$). There weren't significant differences in VEGF levels between patients with FVC <80% ($n = 11$) and those with FVC ≥80% (219.15 ± 252.57 vs 154.26 ± 208.91 pg/ml, respectively).

Mean VEGFR2 levels were also increased in SSc pts compared to controls (5784.6 ± 4773.8 and 1552.6 ± 272.8 pg/ml, respectively; $p < 0.0001$). There weren't any differences between pts with diffuse or limited SSc. In pts with digital ulcers, or normal levels of sPAP, or DLCO ≥50% mean VEGFR2 levels didn't differ from those without digital ulcers, or sPAP >30 mmHg, or DLCO <50%, respectively. Serum VEGF levels directly correlated with sPAP ($R = 0.40$; $p = 0.007$).

Conclusions: Our findings show significantly higher circulating levels of VEGF and VEGFR2 in SSc patients. Associations of VEGF with some clinical signs indicate its role in pathology of SSc.

Disclosure of Interest: None declared

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AB0799

ALTERATIONS OF BODY COMPOSITION IN SCLERODERMA PATIENTS ARE ASSOCIATED WITH DISEASE ACTIVITY AND PHYSICAL ACTIVITY BUT NOT WITH LUNG INVOLVEMENT

S. Oreska¹, M. Spiritovic^{1,2}, P. Cesak², M. Cesak², H. Storkanova¹, H. Smucrova¹, B. Hermankova^{1,2}, O. Ruzikova¹, H. Mann¹, K. Pavelka¹, L. Senolt¹, J. Vencovsky¹, R. Becvar¹, M. Tomcik¹. ¹Department of Rheumatology, 1st Medical Faculty, Charles University, Institute of Rheumatology; ²Department of Physiotherapy, Faculty of Physical Education and Sport, Charles University, Prague, Czech Republic

Background: Systemic sclerosis (SSc) is characterised by fibrosis of the skin and visceral organs, especially digestive tract, and musculoskeletal and lung involvement, which limit mobility/self-sufficiency of patients and can have a negative impact on body composition and physical activity.

Objectives: To assess body composition and physical activity of SSc patients and healthy controls (HC).

Methods: 59 patients with SSc [50 females; mean age 52.5; disease duration 6.7 years; limited cutaneous (lcSSc, 34)/diffuse cutaneous (dcSSc, 25)] and 59 age-/sex-matched HC (50 females, mean age 52.5) without rheumatic diseases were