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ABSTRACT SUPPLEMENT

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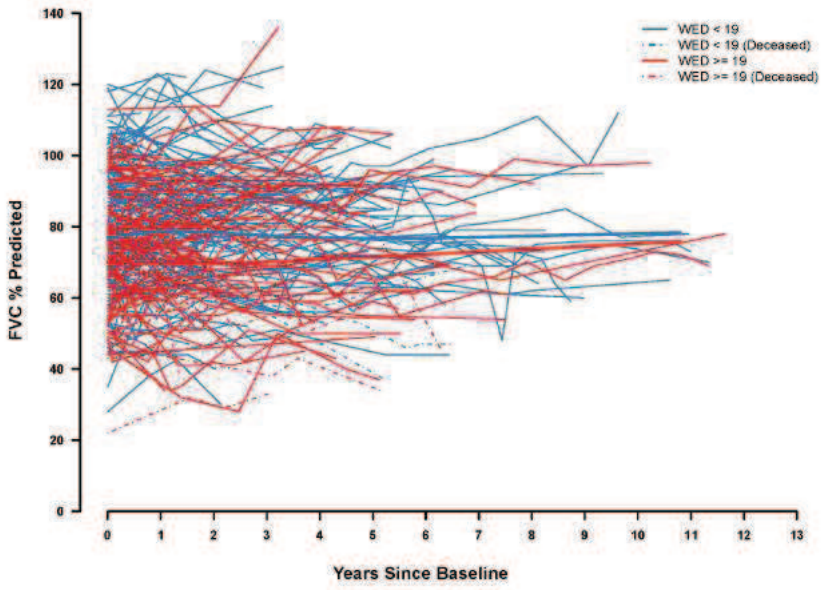
Table 1. Baseline Characteristics (n=249)				
Mean (SD) or n (%)	Total (N=249)	WED <19 (N=142)	WED≥19 (N=107)	p-value
Age at time of HRCT, years	50.9 (11.4)	49.8 (11.4)	52.3 (11.2)	0.084
Sex, women	206 (83%)	130 (92%)	76 (71%)	**<0.001**
Body mass index, kg/m^2	26.2 (5.8)	26.3 (5.3)	26.2 (5.8)	0.933
Ethnicity, white	190 (79%)	111 (80%)	79 (76%)	0.428
Smoker, current or former	93 (38%)	51 (36%)	42 (40%)	0.609
Proton Pump Inhibitor use, baseline	145 (60%)	70 (51%)	75 (73%)	**0.001**
Alcohol, current	103 (52%)	61 (57%)	42 (45%)	0.094
SSc disease subtype, diffuse cutaneous	100 (40%)	52 (37%)	48 (45%)	0.402
Years since first non-Raynaud Symptom	6.6 (10.5)	5.6 (7.4)	7.8 (13.4)	0.129
SSc-specific antibodies, positive	156 (78%)	92 (81%)	64 (75%)	0.359
Anti-topoisomerase I (Scl-70)	72 (34%)	40 (34%)	32 (34%)	0.982
Anti-centromere (ACA)	46 (21%)	25 (20%)	21 (23%)	0.658
RNA polymerase III	43 (25%)	30 (31%)	13 (18%)	0.063
Modified Rodnan skin score	11.4 (10.5)	10.4 (9.3)	12.7 (11.2)	0.107
Baseline radiographic ILD present	182 (73%)	94 (66%)	88 (82%)	**0.005**

Abbreviations: HRCT, high-resolution computed tomography; WED, widest esophageal diameter (median=17mm, range 0-44mm); SSc, systemic sclerosis

1997;169:977-83

Figure 1. Change in Forced Vital Capacity (FVC % Predicted) Over Time in 249 Patients with Systemic Sclerosis



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Abstract Number: 1885

Can Nailfold Videocapillaroscopy Images be Interpreted Reliably By Different Observers? Results of an Inter-Reader and Intra-Reader Exercise Among Rheumatologists with Different Experience in This Field

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SESSION INFORMATION

Session Date: Monday, November 14, 2016
Session Title: Systemic Sclerosis, Fibrosing Syndromes, and Raynaud's – Clinical Aspects and Therapeutics - Poster II
Session Type: ACR Poster Session B
Session Time: 9:00AM-11:00AM

Background/Purpose: Nailfold videocapillaroscopy (VCP) has become an established method to assess the microcirculation in patients with Raynaud's phenomenon and connective tissue diseases. The 2013 Systemic Sclerosis (SSc) criteria included capillaroscopy as a necessary tool for SSc classification. Adequate training for identification of Systemic Sclerosis capillaroscopic patterns is relevant for all Rheumatologists. To date, there is little evidence of the reliability of VCP findings amongst different readers. We evaluated inter- and intra-reader agreement of 13 Rheumatologists to identify SSc capillaroscopy patterns ("early", "active" and "late") proposed by Cutolo et al (ref 1).

Methods: Thirteen rheumatologists (7 without experience and 6 with more than 2 years of experience in the routine performance and reading of capillaroscopic images) received a 20 min training regarding the identification of SSc capillarosocpy patterns to standardize reading criteria. Then, they individually rated 60 videocapillaroscopy images (12 from healthy subjects, 48 from SSc patients) at baseline, and 4 weeks after the first reading using an electronic platform in order to perform the intra-reader exercise. The reading of an expert with more than 15 years of experience in capillaroscopy was considered the gold standard. Data was analyzed using Cohen´s kappa for concordance, Student´s t test and ANOVA were used to compare kappa means for inter-reader, intra-reader and inter-pattern readings.

Results: Mean inter-reader and intra-reader kappa were 0.45 and 0.49, respectively, reflecting moderate agreement. Mean kappa scores were significantly higher among experienced readers when compared with unexperienced readers (inter-reader kappa: 0.58 vs 0.34, p=0.001, intra-reader kappa: 0.65 vs 0.37, p=0.01). Agreement was substantial (kappa =0.61) for the identification of normal vs abnormal capillaroscopy, and higher than the overall agreement (p=0.009). Agreement was higher for the identification of "active" (0.48, p=0.009) and "late" SSc patterns (0.56, p=0.008) than for the identification of "early" SSc pattern (0.35, p=0.003) when compared to overall agreement in all participants. Agreement for "early" and "active" patterns was higher in experienced vs not experienced readers ("early" pattern kappa=0.45 vs 0.26, p=0.01, "active" pattern kappa= 0.62 vs 0.35, p=0.006, "late" pattern kappa=0.66 vs 0.48, p=0.12).

Conclusion: There is moderate agreement among rheumatologists for the identification of SSc videocapillaroscopy patterns, while there is substantial agreement among rheumatologists regardless their experience in videocapillaroscopy, in the identification of normal and abnormal capillaroscopic images. Agreement for the identification of "active" and "late" patterns is higher than for "early" capillaroscopic pattern. The identification of "early" capillaroscopic changes may require more experience in the performance and interpretation of this technique. Ref 1. Cutolo M et al. J Rheumatol 2000;27:155-60.

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Abstract Number: 1886

Right Ventricular Load-Adaptability and Response to Therapy in Scleroderma Versus Idiopathic Pulmonary Arterial Hypertension

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Background/Purpose: Recent studies have suggested that right ventricular (RV) adaptation in patients with scleroderma-associated pulmonary arterial hypertension (SSc-PAH) is worse than in patients with idiopathic pulmonary arterial hypertension (IPAH). This has been proposed as one of the explanations for increased mortality in SSc-PAH. However, few studies have compared incident groups of treatment naive SSc-PAH and IPAH patients. The objective of this study is to compare RV remodeling and load-adaptability metrics in patients with SSc-PAH versus IPAH at the time of diagnostic right heart catheterization (RHC) at our institution and at 1 year follow up.

Methods: A retrospective review of adults with SSc-PAH and IPAH who underwent RHC at Stanford Medical Center between 2002-2016 was performed. Inclusion criteria were as follows: mean pulmonary artery pressure (mPAP) greater than or equal to 25 mmHg, baseline echocardiogram within 3 months of RHC, absence of left heart disease (pulmonary capillary wedge pressure less than or equal to 15 mmHg), and absence of severe ILD (FVC and/or TLC < 70%). Baseline characteristics, RHC parameters, and standard echocardiographic measurements including right ventricular fractional area change (RVFAC), tricuspid annular plane systolic excursion (TAPSE), and RV global longitudinal strain (RV GLS) were obtained. Load-adaptability metrics, including the ratio of cardiac index to mean pulmonary artery pressure (CI/mPAP), right atrial pressure to pulmonary pulse pressure (RAP/PP), and RV global longitudinal strain to pulmonary vascular resistance (RV GLS/PVR) were compared between the SSc-PAH and IPAH groups. We used non-parametric Mann Whitney U-test for continuous variables and Fischers Exact Test for categorical variables. Change in RV function was examined by follow up echo (1 year post-RHC) in a subset of patients matched on PH-directed therapy.