pain in 7(18.4%). Gastrointestinal (GI) symptoms comprised of regurgitation in 31 (81.5%) and dysphagia in 14 (36.8%). Interstitial lung disease was present in 30 (78.9%) patients, with higher prevalence in diffuse scleroderma (100%) than in limited scleroderma (70%) ($p=0.01$). Pulmonary hypertension was present in 18 patients, with significantly higher prevalence in diffuse disease (57.1%), that was secondary to interstitial lung disease and in limited disease it was found in (11.8%) ($p<0.01$). Thirty (78.9%) patients were found to have restricted disease on pulmonary function tests.

Obstetric history showed a higher prevalence of primary infertility in at least 6 (15.8%) patients, with significantly higher prevalence in limited systemic sclerosis disease as compared to diffuse disease (23.5% vs 9.5%, and $p=0.05$). Fibromyalgia diagnosed as per ACR criteria was present in 7 (18.4%) patients, and depression assessed by Hospital Anxiety and depression (HADS) score was present in 10 (26.3%) patients.

Anti nucleic acid antibody (ANA) was found positive in 30 (78.9%) patients. Anti Scl-70 antibodies were in 24 (63.2%) patients, with significant association with diffuse disease (85% vs 35.3% and $p<0.01$), while anti centromere antibodies were present in 20 (52.6%) patients; significantly higher in limited disease (94.2% vs 19.0%, and $p<0.01$).

Conclusion: Scleroderma is a very important, autoimmune multisystem disease. It has female preponderance. Raynaud phenomenon is the most initial clinical feature followed by other manifestations of variable course and disease severity. Interstitial lung disease and pulmonary hypertension were the most important complication found in our patients which has poor prognosis. So, it is imperative to early diagnose and treat the disease manifestations to prevent future complications.

Keywords: Scleroderma, Systemic sclerosis.

DISCLOSURE OF INTEREST:

Sadia Asif: None declared, Muhammad Haroon: None declared, Dr Asadul Khan: None declared, Dr. Muhammad Faiq: None declared DOI: 10.1136/annrheumdis-2021-eular.1635

AB0094

INCREASE OF ENDOTHELIAL PROGENITOR CELLS IN SYSTEMIC SCLEROSIS-ASSOCIATED INTERSTITIAL LUNG DISEASE

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Background: Endothelial progenitor cells (EPC), involved in vasculogenesis and endothelial tissue repair, have been described as relevant players in vascular and connective tissue diseases [1-2]. In this regard, a previous study of our group disclosed that the degree of EPC frequency may help to identify the presence of interstitial lung disease (ILD) in rheumatoid arthritis patients [3]. Given that ILD is the main cause of mortality in patients with systemic sclerosis (SSc) [1, 4-6], the understanding of the role of EPC in the mechanism of SSc-ILD⁺ vasculopathy is crucial.

Objectives: To assess the potential role of EPC on vascular dysfunction associated with the presence of ILD in patients with SSc.

Methods: Peripheral venous blood was collected from a total of 39 patients with SSc, 20 with ILD (SSc-ILD⁺) and 19 without ILD (SSc-ILD⁻). All subjects were recruited from the Rheumatology and Pneumology departments of Hospital Universitario Marqués de Valdecilla, Santander, Spain. Quantification of EPC was analyzed by flow cytometry. EPC were considered as CD34⁺, CD45^{Low}, CD309⁺ and CD133⁺.

Results: Statistically significant differences in EPC frequency between patients with SSc-ILD⁺ and patients with SSc-ILD⁻ were disclosed. Specifically, an increase of EPC frequency was observed in SSc-ILD⁺ patients when compared to patients with SSc-ILD⁻ (mean $\pm$ standard deviation: 0.033 ± 0.012 versus 0.021 ± 0.017 , respectively, $p=0.012$).

Conclusion: Our results suggest a potential role of EPC on vascular damage associated with the manifestation of ILD in patients with SSc.

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Acknowledgements: Personal funds, VP-C: PREVAL18/01 (IDIVAL); SR-M: RD16/0012/0009 (ISCIII-ERDF); LL-G: INNVAL20/06 (IDIVAL); RP-F: START PROJECT (FOREUM); RL-M: Miguel Servet type I CP16/00033 (ISCIII-ESF).

Disclosure of Interests: Verónica Pulito-Cueto: None declared, Sara Remuzgo Martinez: None declared, Fernanda Genre: None declared, Belén Atienza-Mateo: None declared, Victor Manuel Mora-Cuesta: None declared, David Iturbe-Fernández: None declared, Leticia Lera-Gómez: None declared, Raquel Pérez-Fernández: None declared, Diana Prieto-Peña: None declared, Virginia Portilla: None declared, Ricardo Blanco Speakers bureau: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen and MSD, Consultant of: Abbvie, Pfizer, Roche, Bristol-Myers, Janssen and MSD, Grant/research support from: Abbvie, MSD and Roche, Alfonso Corrales: None declared, Jose Manuel Cifrián-Martínez: None declared, Raquel López-Mejías: None declared, Miguel A González-Gay Speakers bureau: Pfizer, Abbvie, MSD, Grant/research support from: Pfizer, Abbvie, MSD

DOI: 10.1136/annrheumdis-2021-eular.2350

AB0095

EXPRESSION AND PATHOGENIC ROLES OF INTEGRIN FAMILY GENE IN SYSTEMIC SCLEROSIS

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Background: Emerging evidence have shown that some integrin members are associated with inflammation and fibrosis in systemic sclerosis (SSc) patients^[1-2]. However, the expression patterns and pathogenic significance of the whole integrin family in SSc are still unclear.

Objectives: This study aimed at evaluating the integrin family gene expression in skin lesion from SSc patients and exploring its potential pathogenic mechanism.

Methods: We utilized the public datasets of SSc skin tissue from Gene Expression Omnibus (GEO) database to analyze the expression and clinical significance of integrin family genes in SSc. In addition, functional enrichment and pathway analysis were also conducted.

Results: Compared with healthy controls, ITGA5, ITGA7, ITGA8, ITGB2, ITGB5, ITGAE and ITGB3BP were abnormally overexpressed in the skin of SSc. Further analysis indicated that ITGA5, ITGA7, ITGA8, ITGB2 and ITGB5 were positively correlated with modified Rodnan skin thickness score (mRSS), while ITGAE and ITGB3BP were negatively correlated with mRSS in SSc. Increased ITGB5 expression was associated with positive of anti-centromere antibody (ACA). Functional enrichment and pathway analysis showed that integrin members had multiple functions in SSc. Among them, ITGA5, ITGB2 and ITGB5 might synergistically promote SSc through affecting extracellular matrix (ECM) turn over, ECM-receptor interaction, focal adhesion and leukocyte trans-endothelial migration. ITGA5 and ITGB5 also affected angiogenesis and endothelial cell function. In addition, ITGA5 was uniquely enriched for actin organization, ITGB5 was uniquely enriched for TGF- β signaling, and ITGB2 was uniquely associated with immune cells activation.

Conclusion: Our results implied that integrins, especially ITGA5, ITGB5, ITGB2 participated in the process of inflammation, vasculopathy and fibrosis in SSc. Together, they might render important therapeutic targets for SSc.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2021-eular.3646

Vasculitis - aetiology, pathogenesis and animal models

AB0096

IGA VASCULITIS AND IGA NEPHROPATHY SHARE A SIMILAR IL17A ASSOCIATION PATTERN

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