with positivity for anti-dsDNA antibodies. A second cohort of 62 SLE patients was included, of which endothelial dysfunction, lipid profile, the presence of atheroma plaques (identified by a pathologic increase in the carotid intimae media thickness -CIMT-), and the frequencies of anti-dsDNA positivity for 7 years, were evaluated. Serum inflammatory and oxidative stress biomolecules, and NETosis-derived bioproducts were further evaluated by multiplex assay and specific commercial kits, respectively. Besides, miRNnomes were identified using next-generation sequencing. Clinical significance of the biomolecules analyzed was explored by correlation/association studies with immunological and CV-risk features.

Results: A significant relationship among the incidence of CVD (i.e. thrombosis or cardiac involvement) and the positivity for anti-dsDNA antibodies was recognized in the first SLE cohort. Accordingly, in the second SLE cohort, significantly impaired micro-vascular endothelial function (identified by reduction of hyperemia post-occlusion area), increased atherogenic index and pathologic increase in the CIMT were assessed in patients positive for anti-dsDNA in relation to anti-dsDNA negative patients. Around a 65% of SLE patients displayed a sustained positivity for anti-dsDNA antibodies for more than 7 years. These patients showed a distinctive and specific molecular profile compared with patients that had remained negative for anti-dsDNA, including increased inflammatory profile (IL1B, IL2, IL6, IL17, EOTAXIN, FGF, GMCSF, IFNγ, IP10, RANTES, TNF), enhanced oxidative status (lipoperoxides), and higher NETosis (nucleosomes, elastase). Levels of those biomolecules were closely interconnected and associated to their regulatory miRNAs, which accordingly exhibited differential expression in SLE anti-dsDNA(+) *vs* anti-dsDNA(-) patients. Finally, the frequency for positivity of anti-dsDNA significantly correlated both with markers of endothelial dysfunction and with the presence of atheroma plaques in SLE patients, pointing at the direct involvement of anti-dsDNA-Abs in the development of these processes.

Conclusion: 1. Positivity for anti-dsDNA antibodies confers a specific inflammatory/oxidative profile linked to an enhanced CV-risk in SLE patients. 2. Moreover, the sustained positivity for anti-dsDNA antibodies fosters the establishment of an atherothrombotic status in these autoimmune patients.

Acknowledgments: Supported by the EU/EFPIA –IMI-JU PRECISESADS (n° 115565) and ISCIII (PI18/0837 and RIER RD16/0012/0015), Co-funded with FEDER.

Disclosure of Interests: Inmaculada Concepcion Aranda-Valera: None declared, Alejandra M. Patiño-Trives: None declared, Roldán Molina Rosa: None declared, Maria A Aguirre: None declared, Pérez Sánchez Laura: None declared, Carlos Pérez Sánchez: None declared, María Luque-Tévar: None declared, Iván Arias de la Rosa: None declared, Maria del Carmen Abalos-Aguilera: None declared, Desiree Ruíz-Vilchez: None declared, Mario Espinosa: None declared, Nuria Barbarroja Puerto Grant/research support from: ROCHE and Pfizer., Speakers bureau: ROCHE and Celgene., Eduardo Collantes-Estévez Grant/research support from: ROCHE and Pfizer., Speakers bureau: ROCHE, Lilly, Bristol and Celgene., Chary Lopez-Pedrera Grant/research support from: ROCHE and Pfizer.

DOI: 10.1136/annrheumdis-2020-eular.4923

FRIDAY, 05 JUNE 2020

Paediatric rheumatology

FRI0454 UNDER DETECTION OF INTERSTITIAL LUNG DISEASE IN JUVENILE SYSTEMIC SCLEROSIS (JSSC) UTILIZING PULMONARY FUNCTION TESTS. RESULTS FROM THE JUVENILE SCLERODERMA INCEPTION COHORT

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Background: Juvenile systemic sclerosis (jSSc) is an orphan disease with a prevalence in around 3 in a million children. Pulmonary involvement in jSSc occurs in approximately 40 % in the inception cohort. Traditionally in jSSc, pulmonary function testing (PFT) with FVC and DLCO are used for screening and computed tomography (HRCT) was more reserved for those with abnormal PFTs. More recently, it has become apparent that PFTs might not be sensitive enough for detecting ILD in children.

Objectives: Utilizing a prospective international juvenile systemic scleroderma cohort (JSScC) [2], to determine if pulmonary screening with FVC and DLCO is sufficient enough to assess the presence of interstitial lung disease in comparison to CT evaluation.

Methods: The international juvenile systemic scleroderma cohort database was queried for available patients with recorded PFT parameters and HRCT performed to determine sensitivity of PFTs detecting disease process.

Results: Of 129 patients in the jSScC, 67 patients had both CT imaging and an FVC reading from PFTs for direct comparison. DLCO readings were also captured but not in as many patients with tandem HRCT (n =55 DCLO and HRCT scan). Therefore, initial analyses focused on the sensitivity, specificity and accuracy of the FVC value from the PFTs to capture the diagnosis of interstitial lung disease as determined by HRCT.

Overall, 49% of the patients had ILD determined by HRCT, with 60% of patients having normal FVC (>80%) with positive HRCT findings, and 24% of patients having normal DLCO (> 80%) with positive HRCT findings. Fourteen percent (n = 3/21) of patients with both FVC and DLCO values within the normal range had a positive HRCT finding.

Conclusion: The sensitivity of the FVC in the JSScC cohort in detecting ILD was only 39%. Relying on PFTs alone for screening for ILD in juvenile systemic sclerosis would have missed the detection of ILD in almost 2/3 of the sample cohort, supporting the use of HRCT for detection of ILD in children with SSc. In addition, the cut off utilized, of less than 80% of predicted FVC or DLCO could be too low for pediatric patients to exclude beginning ILD. This pilot data needs confirmation in a larger patient population.

Supported by the “Joachim Herz Stiftung”

Disclosure of Interests: Ivan Foeldvari Consultant of: Novartis, Bernd Hinrichs: None declared, Kathryn Torok: None declared, Maria Jose Santos Speakers bureau: Novartis and Pfizer, Ozgur Kasapcopur: None declared, Amra Adrovic: None declared, Valda Stanevicha: None declared, Flávio R. Sztajnbok: None declared, Maria T. Terreri: None declared, Ana Paula Sakamoto: None declared, Ekaterina Alexeeva Grant/research support from: Roche, Pfizer, Centocor, Novartis, Speakers bureau: Roche, Novartis, Pfizer., Jordi Anton Grant/research support from: grants from Pfizer, abbvie, Novartis, Sobi. Gebro, Roche, Novimmune, Sanofi, Lilly, Amgen, Grant/research support from: Pfizer, abbvie, Novartis, Sobi. Gebro, Roche, Novimmune, Sanofi, Lilly, Amgen, Consultant of: Novartis, Sobi, Pfizer, abbvie, Consultant of: Novartis, Sobi, Pfizer, abbvie, Speakers bureau: abbvie, Pfizer, Roche, Novartis, Sobi, Gebro, Speakers bureau: abbvie, Pfizer, Roche, Novartis, Sobi, Gebro, Maria Katsikas: None declared, Vanessa Smith Grant/research support from: The affiliated company received grants from Research Foundation - Flanders (FWO), Belgian Fund for Scientific Research in Rheumatic diseases (FWRO), Boehringer Ingelheim Pharma GmbH & Co and Janssen-Cilag NV, Consultant of: Boehringer-Ingelheim Pharma GmbH & Co, Speakers bureau: Actelion Pharmaceuticals Ltd, Boehringer-Ingelheim Pharma GmbH & Co and UCB Biopharma Sprl, Rolando Cimaz: None declared, Mikhail Kostik: None declared, Simone Appenzeller: None declared, Mahesh Janarthanan: None declared, Monika Moll: None declared, Dana Nemcova: None declared, Dieneke Schonenberg: None declared, Cristina Battagliotti: None declared, Lillemor Berntson Consultant of: paid by Abbvie as a consultant, Speakers bureau: paid by Abbvie for giving speaches about JIA, Blanca Bica: None declared, Juergen Brunner Grant/research support from: Pfizer, Novartis, Consultant of: Pfizer, Novartis, Abbvie, Roche, BMS, Speakers bureau: Pfizer, Novartis, Abbvie, Roche, BMS, Patricia Costa Reis: None declared, Despina Eleftheriou: None declared, Liora Harel: None declared, Gerd Horneff Grant/research support from: AbbVie, Chugai, Merck Sharp & Dohme, Novartis, Pfizer, Roche, Speakers bureau: AbbVie, Bayer, Chugai, Merck Sharp & Dohme, Novartis, Pfizer, Roche, Dragana Lazarevic: None declared, Kirsten Minden Consultant of: GlaxoSmithKline, Sanofi, Speakers bureau: Roche, Susan Nielsen: None declared, Farzana Nuruzzaman: None declared, Anjali Patwardhan: None declared, Yosef Uziel: None declared, Nicola Helmus: None declared

DOI: 10.1136/annrheumdis-2020-eular.1788

FRI0455 IS THERE AN INCREASE IN THE FREQUENCY OF INFLAMMATORY DISEASES IN THE FAMILIES OF PATIENTS WITH FMF?

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Background: Familial Mediterranean Fever (FMF) is the most common periodic fever syndrome in childhood with an autosomal recessive inheritance pattern and is characterized by unprovoked fever attacks, serositis episodes. The causative gene of the disease is MEFV that encodes pyrin protein. The pyrin protein takes a role in pathways related to inflammation, and mutations of it lead to increased inflammation. It is already shown that frequencies of some certain diseases like PAN, HSP increase in patients with FMF. Nevertheless,