**Background:** Sjögren's syndrome (SS) affects mainly individuals of the 4th or 5th decade of life, although patients with early (≤35 years old) or late (≥65 years old) disease onset have been described in the literature. The clinical spectrum of the disease extends from mild dryness to severe systemic vasculitis and lymphoproliferative disorders. The phenotypic diversity of SS is defined by many factors, including age, since many parameters related to age may affect the clinical expression of the disease. Few studies have been conducted to study the effect of age on the clinical phenotype of SS, though with limited number of patients. Large and well-defined groups of SS are required to address such questions. Objectives: To study the clinical phenotype of SS patients with early and late disease onset and to explore the association of age with lymphoma development in a unified multicenter cohort.

**Methods:** From a total cohort of 1979 consecutive SS patients who fulfilled the 2016 EULAR/ACR criteria and are followed up in 5 clinical centers (Università of Udine, Pisa and Athens, Harokopio and Ioannina, (UAPHI)), those with either early (≤35 years) or late (≥65 years) disease onset were identified and matched according to gender and disease duration with middle aged controls whose disease onset was at the 4th or 5th decade of life. Glandular manifestations, extra-glandular manifestations, serologic characteristics and histologic features were compared between the 2 age groups and the middle-aged control groups. Statistical analysis for categorical variables was performed by Fisher exact or chi-square tests and for continuous variables with t test or Mann-Whitney accordingly.

**Results:** Three hundred seventy-nine (19%) SS patients with early and 293 (15%) with late disease onset were identified and compared with 353 and 285 middle aged SS controls respectively. The median disease duration of patients with early onset was 12 years (range:0-68) and for those with late disease onset was 5 years (range: 0-27). SS patients with early disease onset had statistically significant higher frequency of Raynaud's phenomenon, lymphadenopathy, hyperechoic bands on parotid-Sub-SSA, anti-SphK1, rheumatoid factor, salivary gland enlargement, low C4 complement levels, leukopenia and lymphopenia (10.3% vs 5.7%, p=0.03, OR=1.91, 95% CI: 1.11-3.27) while SS patients with late disease onset had more frequently dry mouth, intestinal lymph disease and lymphoma (6.8% vs 2.1%, p=0.04, OR=3.4, 95% CI: 1.35-8.1).

**Conclusion:** In a multicenter cohort of 1979 consecutive SS patients, those with early and late disease onset comprise more than one third of the total SS population. Patients with early disease onset, exhibit robust B cell factors with traditional risk factors for lymphoma as opposed to patients with late disease onset. Both age groups have increased lymphoma prevalence but presumably for different reasons, since late onset patients lack classical predictors of lymphoma. Therefore, these predictors deserve further study in different disease subsets.

**Disclosure of Interests:** Andreas Goulis: None declared, Ourania Argyropoulou: None declared, Saviana Gandolfo: None declared, Valentina Donat: None declared, Marco Binutti: None declared, Sara Zandonella Callegher: None declared, Loukas Chatzis: None declared, Alki Venetsanopoulou: None declared, Evangelia Zampeli: None declared, Maria Mavromati: None declared, Paraskevi Vougli: None declared, Clio Mavragani: None declared, Chiara Baldini: None declared, Fatima Sokok: None declared, Dimitris Fotiadis: None declared, Silvia Fonzetti: None declared.

**Disclosure:** Haralampos M. Moutsopoulos: None declared, Athanasios Tzioufas: None declared, Maria Loukianou: None declared, Clio Mavragani: None declared, Chiara Baldini: None declared, Paraskevi Vougli: None declared, Antonella Cecchettini: None declared, Chiara Baldini: None declared. DOI: 10.1136/annrheumdis-2020-eular.991
Conclusion: Cardiovascular, bone and neurologic comorbidities are frequently detected already at the time of diagnosing SLE. High numbers of medical prescriptions and hospitalization following SLE diagnosis reflect the comprehensive disease burden. Differences to controls without autoimmune disease are overestimated by detection already at the time of diagnosing SLE. High numbers of medical prescriptions and hospitalization following SLE diagnosis reflect the comprehensive disease burden. Differences to controls without autoimmune disease are overestimated by detection already at the time of diagnosing SLE.

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Background: Major salivary gland ultrasonography has an established role in diagnosis and assessment of pSS. Nowadays, however, interest is also growing in using a higher resolution ultrasound (UHFUS) technique, which can produce frequencies up to 70 MHz and achieve tissue resolution up to 30 μm, opening up new possibilities for the study of salivary glands.

Objectives: To investigate the usefulness of UHFUS in LSG ultrasound-guided biopsy and preoperative planning.

Methods: Consecutive patients undergoing LSG for clinically suspected pSS were included in this study from January 2018 to December 2019. UHFUS of LSG was performed by using VEVO MD, equipped with a 70 MHz probe, scanning first the central compartment of the inferior lip, and then both peripheral compartments. The following parameters were evaluated: distribution of the glands, parenchymal inhomogeneity (score 0-3, from normal to evident), and fibrosis. UHFUS imaging was used to help locate the LSG for the US-guided biopsy. The same expert pathologist calculated the surface area of gland sections examined, the LSG focus score (FS), the number of foci and evaluated the presence of ectopic germinal centers (GCs). Consecutive patients that had undergone a traditional LSG biopsy from December 2016 to December 2017 were included as controls.

Results: We included a total of 249 patients with suspected pSS: 137 undergoing the UHFUS-guided LSGs and 112 the traditional LSG biopsy procedure. No demographic differences were observed between the two groups. No differences were also observed in the distribution of the final diagnosis. A diagnosis of pSS according to the ACR 2016 criteria was made in 60/137 (43.8%) and 36/112 (32.1%) patients, respectively whereas a diagnosis of no-SS sicca was made in 44/137 (32.1%) and in 43/112 (38.4%) patients; the remaining diagnosis included secondary SS (4/137, 3% and 9/112, 8%) and undifferentiated connective tissue disease (UCTD) (29/137, 21.2%, and 24/112, 21.4%). With respect to no-SS sicca controls and UCTD patients, pSS patients presented higher UHFUS inhomogeneity scores in both central and peripheral labial compartments (p=0.001). There were no complications from the HUFUS-guided LSG biopsy. The mean glandular surface area obtained was significantly higher than the area obtained in controls (p=0.02) thus facilitating the assessment of the FS. Interestingly, the latter showed a good correlation with the UHFUS inhomogeneity (r=0.509**, p=0.000).

FR1053

ULTRA HIGH-RESOLUTION ULTRASOUND (UHFUS) OF LABIAL SALIVARY GLANDS: POTENTIAL APPLICATIONS IN PRIMARY SJÖGREN’S SYNDROME


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Background: Major salivary gland ultrasonography has an established role in diagnosis and assessment of pSS. Nowadays, however, interest is also growing in using a higher resolution ultrasound (UHFUS) technique, which can produce frequencies up to 70 MHz and achieve tissue resolution up to 30 μm, opening up new possibilities for the study of salivary glands (LSG).

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Methods: Consecutive patients undergoing LSG for clinically suspected pSS were included in this study from January 2018 to December 2019. UHFUS of LSG was performed by using VEVO MD, equipped with a 70 MHz probe, scanning first the central compartment of the inferior lip, and then both peripheral compartments. The following parameters were evaluated: distribution of the glands, parenchymal inhomogeneity (score 0-3, from normal to evident), and fibrosis. UHFUS imaging was used to help locate the LSG for the US-guided biopsy. The same expert pathologist calculated the surface area of gland sections examined, the LSG focus score (FS), the number of foci and evaluated the presence of ectopic germinal centers (GCs). Consecutive patients that had undergone a traditional LSG biopsy from December 2016 to December 2017 were included as controls.

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FR1052

PULMONARY HYPERTENSION IN NEWLY DIAGNOSED SPANISH PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS: DATA FROM THE RELES COHORT


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Objective: To investigate the incidence of pulmonary hypertension (PH) in 54 Spanish patients with systemic lupus erythematosus (SLE) newly diagnosed and treated at 8 hospitals in Spain.

Methods: A prospective observational study on a multicenter Spanish inception cohort. Patients with SLE, diagnosed by the American College of Rheumatology (ACR) criteria, since January 2009, who had at least one transthoracic echocardiogram (TTE) performed were selected. Demographic data, diagnostic criteria, follow-ups, treatments and SLEDAI were analyzed.

Results: Of 289 patients diagnosed with SLE with TTE performed, 15 (5.2%) patients were identified to have PH. Mean age was 56.9±7.7 years, of which 93.3% (14) were women and 80% (12) Caucasian. The ACR score at diagnosis was 4.66. Mean SLEDAI was 15. Only 5 patients had dyspnea at the time of diagnosis. Mean pulmonary arterial systolic pressure was 49.2±5.6 mmHg. Among the PH, 4 patients had pericarditis (26.6%), 3 (20%) valvulopathies (1 antiphospholipid syndrome), 1 patient pulmonary embolism and 1 shrinking lung. Multivariable analysis indicated that pericarditis (odds ratio (OR)=2.53), and valvulopathies (OR 8.96) were independently associated with the development of PH in SLE. Having PH was associated with older age at diagnosis (p<0.001), more dyspnea (p<0.001), higher ESR (p=0.007), more serositis (p<0.001), higher SLEDAI (p=0.011), higher SLICC (p<0.001), higher number of admissions (p=0.006) and higher mortality (p=0.003).

Conclusion: PH in SLE is a serious comorbidity with high mortality. In the RELES cohort it was associated with increased disease activity, pericarditis and valvulopathies. Performing TTE in patients with SLE may favor early diagnosis and treatment.

References:

Disclosure of Interests: Jorge Álvarez Troncoso: None declared, Ángel Robles Marhuenda: None declared, Francesca Mitjavila Villero: None declared, Francisco José García Hernández: None declared, Adela Marín Balvé: None declared, Antoni Castro Consultant of: Actelion pharmaceuticals, GSK, MSD., Gonzalo Salvador Cervelló: None declared, Eva Fonseca: None declared, Isabel Perales Fraile: None declared, Guillermo Ruiz-Irastorza: None declared

DOI:10.1136/annrheumdis-2020-eular.991