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Objectives: Baseline characterization of European patients diagnosed with primary Sjögren syndrome (SS) according to the 2002 AE criteria.

Methods: The Big Data Sjögren Project was formed in 2014 to take a "high-definition" picture of the main features of primary SS by merging international SS databases. International experts of the EULAR-SS Task Force were invited to participate. By January 2017, the database included 9302 consecutive patients recruited from 21 countries of the 5 continents.

Results: A total of 6586 (71%) patients were included from European countries. In comparison with non-European countries, European patients had a higher mean age (54 v 51 yrs, $p<0.001$), higher frequency of men (7% v 5%, $p=0.001$), dry eyes (94% vs 88%, $p<0.001$), dry mouth (94% vs 91%, $p<0.001$), and lower frequency of abnormal ocular (84 vs 86%, $p=0.049$) and oral (75 vs 81%, $p<0.001$) tests. Immunologically, European patients had a lower frequency of anti-Ro/La antibodies (69 vs 78%, $p<0.001$) and a higher frequency of RF (50 vs 47%, $p=0.01$), low C4 (14 vs 9%, $p<0.001$) and cryoglobulins (8% vs 3%, $p<0.001$). Logistic regression identified as independent variables older age (OR 1.02), male gender (OR 2.62), abnormal oral tests (OR 0.26), anti-Ro/La antibodies (OR 0.69), RF (OR 1.76), low C4 (OR 1.97) and cryoglobulins (OR 3.85).

Conclusions: European patients are diagnosed at older age, are more frequently men, and presented a lower frequency of anti-Ro/La antibodies and a higher frequency of immunological markers related to mixed cryoglobulinemia.

Disclosure of Interest: None declared

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SAT0303 CLINICAL DIFFERENCES BETWEEN DEFINITE AND PROBABLE ANTIPHOSPHOLIPID (APS) PATIENTS: SHOULD THEY BE TREATED THE SAME?

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Background: The management of patients with recurrent thromboses/pregnancy morbidity and transient detection of antiphospholipid antibodies (aPL) can be a medical challenge. Although these patients are commonly seen in practice, there are no specific guidelines for the treatment in this situations.

Objectives: To investigate the clinical differences between definite and probable APS patients.

Methods: We performed a cross-sectional study in a group of 90 outpatients seen in our department. Seventy-seven of them met the Sydney classification criteria for definite APS, and thirteen had thrombosis or gestational morbidity, but no definite serological criteria for the diagnosis of APS. Clinical and serological features were collected during visits and by chart review, and the two groups were compared. Transient aPL was defined as only one detection of aPL (lupus anticoagulant, anticardiolipin IgM/IgG and/or anti-beta-2-glycoprotein 1 IgM/IgG) after 2 or more assays, at least 12 weeks apart.

Results: Demographic and clinical characteristics are shown in Table 1. In a bivariate analysis, there was no difference between groups regarding the criteria and non-criteria manifestations of APS, except for the presence of livedo ($p=0.033$). In a multivariate regression analysis, the model was adjusted to age, sex, and variables with $p<0.10$ in the bivariate analysis (age, sex, race, livedo,

Table 1. Demographic and clinical characteristics (N=90)

Variable	Definite APS (N=77)	Probable APS (N=13)	P value
Age	42.0±12	40.5±10.3	NS
Female gender	64 (83.1)	12 (92.3)	NS
Caucasian	50 (64.9)	8 (61.5)	NS
Time first manifestation (mo)	120 (33.3–50)	174 (81–202)	NS
Criteria manifestations			
Thrombotic	72 (93.5)	13 (100)	NS
Obstetric	28 (43.8)*	1 (8.3)**	NS
Non criteria			
Livedo	20 (26)	0 (0)	$p=0.033$
Thrombocytopenia	11 (14.3)	0 (0)	NS
Valvulopathy**	7 (11.7)†	1 (11.1)**	NS
Raynaud phenomenon	21 (27.3)	1 (7.7)	NS
Migraine	36 (46.8)	8 (61.5)	NS

*N=64; **N=12, †N=60, ‡N=9. Mo = months. CVD = cardiovascular disease. Values showed as N (%) for categorical variables, mean ± SD for normal distribution and Median (interquartile range) for asymmetric distribution.

Raynaud's phenomenon). No difference between groups was found after the analysis.

Conclusions: This study suggests patients with transient detection of aPL have the same clinical characteristics of patients with definite APS, including thrombotic features, pregnancy morbidity, and non-criteria manifestations. In this context, our data suggests that both groups should be treated according to the current treatment guidelines for APS.

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SAT0304 STIMULATORY AND INHIBITORY KILLER IMMUNOGLOBULIN-LIKE RECEPTORS ON NATURAL KILLER T (NKT) CELLS IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE): RELATION TO DISEASE ACTIVITY

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Background: Natural killer T (NKT) cells are a unique subgroup of T cells that represents a bridge between innate and adaptive immunity. The functions of NKT-cells are regulated by the balance between activating and inhibitory Killer cell immunoglobulin-like receptors (KIRs). Systemic lupus erythematosus (SLE) patients showed aberrant expression of KIRs on NKT-cells. Whether the expression pattern of KIRs on NKT-cells is associated with disease activity in SLE is still unknown.

Objectives: Assessment of expression of stimulatory and inhibitory KIRs on NKT-cells in SLE patients and its relation to disease activity.

Methods: We recruited 40 SLE patients and 20 age and gender matched healthy controls. According to SLE disease activity index (SLEDAI), patients were divided into two groups; active SLE (n=20) and inactive SLE (n=20). SLE was active when SLEDAI was ≥ 4 . Immuno-phenotyping by flow cytometry was done using markers for NKT-cells (CD3 and CD56), stimulatory KIRs (KIR2DL4, CD158D) and inhibitory KIRs (KIR3DL1, CD158E1). Absolute counts and percentage of NKT cells expressing CD158D and CD158E1 together with their mean fluorescence intensity (MFI) were measured. The histogram of CD158 expression was used to assess KIRs on NKT cells.

Flow cytometry charts of lymphocytes and NKT cells in SLE patients (active and inactive) and controls are shown in (Figure 1).

Laboratory work included ANA, anti-dsDNA, Anti-smith, C3, C4, CRP and ESR.

Results: Mean age of patients was 29.9±10.8 years. Females constituted 95% (n=38) of patients. Mean disease duration was 4.4±4.5 years. Mean SLEDAI was 11.75±7.43.

NKT-cells absolute number (cell/mm³) was significantly decreased in SLE patients (89±135) compared to controls (287±133), ($P=0.001$). Likewise, the absolute number of NKT-cells (cell/mm³) was significantly lower in active SLE patients (65.2±57.8) compared to inactive ones (114.5±181.1), ($P=0.001$).

