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Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2017-eular.6619

SAT0286 INCIDENCE AND MORTALITY OF PRIMARY SJÖGREN'S SYNDROME: TIME TRENDS OVER A 40-YEAR PERIOD IN A POPULATION-BASED COHORT IN THE UNITED STATES

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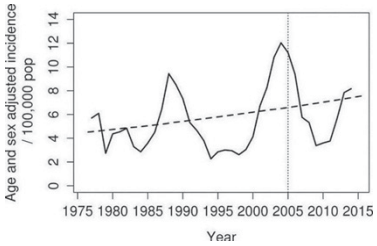
Background:
Few studies have reported the incidence of primary Sjögren's syndrome (pSS) in well-defined populations worldwide, and none of them covered a period of time long-enough to analyze potential time trends in incidence rates. Whether pSS is associated with a higher mortality rate compared to the general population is also unclear from previously published studies.
Objectives: To estimate the incidence and mortality rates of pSS among residents of Olmsted County, Minnesota, and their evolution over time.
Methods: All medical records of patients with a diagnosis or suspicion of SS in Olmsted County, MN, from January 1, 2006 to December 31, 2015 were reviewed to identify incident cases of pSS (defined according to physician diagnosis). All patients with doubtful cases and all patients with an associated systemic autoimmune disease were excluded. These cases were combined with a previous 1976–2005 incident cohort from the same population (reference). Incidence rates were age and sex adjusted to the US white 2010 population. Survival rates were compared with the expected rates in the general population of Minnesota.
Results: With 61 incident cases of pSS diagnosed in Olmsted County in 2006–2015, the total cohort included 172 patients with incident pSS in 1976–2015. Of the 172 patients, 151 (88%) were women and 161 (94%) were white, with a mean (SD) age at diagnosis of 58.3 (16.7) years. The average age- and sex- adjusted annual incidence for 2006–2015 was 5.9 per 100,000 population (95% CI 4.4–7.4), and overall incidence for the entire period was 5.8 (95% CI: 4.9–6.6) per 100,000. Incidence was 2 to 7 times higher in females compared to males in the different age classes (5.9 times higher on average), and increased progressively with age, culminating at 19.6 per 100,000 in females aged 65–74 years, with a slight decline thereafter to 15.9 per 100,000 among females aged 75 years and older. The incidence increased with calendar time over the 40-year period (p=0.005, figure). There was no apparent seasonality in the incidence of pSS, with similar number of cases diagnosed during all four seasons. There was no difference in mortality in the pSS cohort compared to expected (standardized mortality ratio 1.15, 95% CI 0.86–1.50).



Conclusions: The average annual incidence of pSS in this population based-cohort was 5.8/100,000, with a progressive increase over the 40 years of the study. Overall survival of pSS patients was not different from the general population.
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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.1178

SAT0287 TH1, TH2 AND TH17 LYMPHOID SUBPOPULATIONS IN PRIMARY ANTIPHOSPHOLIPID SYNDROME

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Background: Primary antiphospholipid antibody syndrome (PAPS) is characterized by thrombosis at different levels and maternal fetal complications in the presence of antiphospholipid antibodies (aPL). Lymphoid subpopulations and cellular immune responses have not been fully studied.
Objectives: To analyze the lymphoid subpopulations, Th1, Th2 and Th17 immune response in patients with PAPS and long term evolution.
Methods: Patients with PAPS,>18 years of age, of both sexes and a group of healthy blood donors matched for age and sex were included. All patients were receiving oral anticoagulants (Coumadin type). No patient had a recent episode of thrombosis or other manifestation of APS at the time of the study. Peripheral blood was obtained and lymphoid subpopulations were determined by flow cytometry in order to identified with specific immunological markers for Treg cells: CD4+/CD25+/FoxP3+ and CD8+/CD25+/FoxP3+. The dendritic cells analyzed were: Type 1: Lin1- HLA-DR+/CD11c+; Type 2: Lin-HLA-DR+/CD123+; B lymphocytes with antiCD19-APC; Monocytes with anti-CD14-PE; NK: CD3-/CD16+56+ and NKT: CD3+/CD16+56+ lymphocytes. Th1 cells were identified by IFN-g$^+$ positivity; Th2: positivity for IL-4+; Th17: positivity for IL-17+. Parametric statistics and Mann-Whitney U-test were used.
Results:

	PAPS (cells/μL)	Healthy controls (cells/μL)	p
Total leukocytes	5655	7000	<0.05*
Lymphocites	1800	2085	NS
Total CD3	1033	1338	NS
Total CD4	701	775	NS
Total CD8	258	441	<0.05*
Monocytes	213	219	NS
B lymphocites	41	86	NS
NK	118	177	NS
NKT	33	45	NS
iNKT	9	20	<0.005**
Tγδ Lymphocites	34	45	NS
LTreg 4 Lymphocites	23	13	NS
Treg 8 Lymphocytes	26	13	NS
DC1	2	4	<0.005**
DC2	1	5	<0.0005***

A total of 39 patients with PAPS were included, age: 51.9±12.8, evolution time: 12.8±8.9 years and 35 healthy controls. In patients with PAPS there was a decrease in the total CD8 count ($p<0.05$) in iNKT ($p<0.005$), DC1 ($p<0.005$) and DC2 ($p<0.0005$) cells compared to the control group (Table 1).We found significant decrease in Th1, Th2 and Th17 cytokines basal and after activation compared to healthy controls.
Conclusions: This study shows profound alterations in innate and adaptive immunity in patients with long-term PAPS, characterized by a decrease in certain lymphocyte subpopulations, with consequent functional alteration. These abnormalities can become new therapeutic targets in order to restore immune imbalance. Our findings may explain in part, the development of thrombosis and other complications, despite treatment with oral anticoagulants in PAPS patients with long term disease evolution.
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Disclosure of Interest: None declared
DOI: 10.1136/annrheumdis-2017-eular.6222

SAT0288 QUALITY OF LIFE IN PATIENTS WITH SYSTEMIC LUPUS ERIYTHEMATOSUS

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Background: The Quality of life (QoL) in patients with SLE is deeply affected by multiple factors including psychological and physical factors, the functional status and the general perception over health.
Objectives: Evaluation of Quality of life and the perceptions over the disease in patients with SLE.functional status and the general perception over health.
Methods: This is a 10 month prospective, cross-sectional study performed on 52 patients hospitalized diagnosed with SLE according to SLICC 2012 criteria. There were evaluated demographic data, organs manifestations, disease activity scores (SLEDAI, SLICC) and treatment. To evaluate the QoL several questionnaires were performed: Health Assessment Questionnaire (HAQ), EuroQol five dimensions questionnaire (EQ5d), Illness Perception Questionnaire and SF-36.
Results: All patients were women with the mean age 49 years with a mean durations of illness 136 month (14). The mean SLEDAI – 5,52 (5,37) and SLICC – 2,25 (1,71).
The mean HAQ value was 0,83 (0,81). The HAQ score doesn't correlate with SLICC (p=0,461) and with SLEDAI (p=0,172), but it was correlated with the neuropsychiatric manifestations. So the psychiatric affectation influences performing the usually daily activities (p=0,005).
When assessing Illness Perception Questionnaire, patients considered their life seriously affected by pain, mean value 6,3 (2,6), 72% consider the illness will be