Spearman correlation test and the association between MMP9 and serology were evaluated with χ^2 test.

Table 1. Demographic characteristics in Hispanic patients with primary Sjögren's Syndrome

Age, median (DS)	53.23 (13.0)
Female, n (%)	58 (96.7)
Anti-Ro/SSA, n (%)	41 (68.3)
Anti-La/SSB, n (%)	18 (31.6)
RF IgG, n (%)	13 (21.7)
RF IgM, n (%)	31 (51.7)
RF IgA, n (%)	24 (40.0)
Schirmer, n (%)	38 (63.3)
OSDI, n (%)	52 (86.7)
OSS, n (%)	43 (71.7)
Metalloproteinase 9, n (%)	14 (23.3)
Ocular Osmolarity >300, n (%)	45 (75.0)
Abnormal USF, n (%)	45 (75.0)
GSMB, n (%)	51 (85.0)

RF: Rheumatoid Factor, OSDI: Ocular Surface Disease Index, OSS: Ocular Staining Score, USF: Unstimulated Salivary Flow; MSGB: Minor Salivary Gland Biopsy.

Conclusion: A significant correlation was observed between serology, ocular surface damage (OSS), and ocular surface inflammation (MMP-9). The positivity of RF isotypes IgM and IgA could alert the clinician about the ocular damage to initiate a study directed to this organ. Nevertheless this present study shows preliminary outcomes, we need to enroll more patients to obtain better outcomes.

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FRI0221 CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME IN PREGNANCY: CASE SERIES

Nur Azizah Allameen, Aisha Lateef, Anita Yee Nah Lim. *National University Hospital, Singapore, Division of Rheumatology, University Medicine Cluster, Singapore, Singapore*

Background: Catastrophic antiphospholipid syndrome (CAPS) is a rare condition associated with high mortality¹. Prompt recognition and treatment is important for optimal outcomes. Clinical features of CAPS can overlap with obstetric complications, leading to diagnostic challenge².

Objectives: Describe two patients with CAPS during immediate post-partum period.

Methods: We report two cases in a single centre over two years.

Results: A 38 year old with obstetric Antiphospholipid syndrome (APS) presented with epigastric pain, transaminitis and thrombocytopenia in keeping with evolving HELLP syndrome. Spontaneous complete miscarriage ensued. Post-delivery, she developed worsening abdominal pain and transaminitis (Table 1). Imaging demonstrated hepatic infarcts and patchy ground glass lung opacities. She was treated for CAPS with intravenous steroids and anticoagulation, and recovered.

A 39 year old with primary APS (deep venous thrombosis and obstetric features) whose antenatal course was fraught with minor per vaginal bleeding (PVB) presented with heavy PVB at 22 weeks gestation requiring cessation of anticoagulation. She developed a deep venous thrombosis. Risk of clot propagation versus bleeding prompted cautious anticoagulation. Labour was induced for declining maternal health and poor foetal prognosis at 24 weeks, 3 days. Post-delivery, she developed abdominal pain, headache, transaminitis and worsening thrombocytopenia with heavy PVB requiring uterine artery embolization. Given features that were consistent with micro thrombi and ischaemia (Table 1), she was treated as for CAPS and HELLP syndrome with intravenous steroids, plasma exchange (PLEX) and intravenous immunoglobulin (IVIG), and recovered.

Table 1

	Case 1	Case 2
Gestational age on admission	17 weeks 5 days	22 weeks 5 days
Treatment during pregnancy prior to CAPS	LMWH ¹ once daily, Aspirin	LMWH twice daily, Aspirin, Steroids, Hydroxychloroquine, monthly IVIG
Time of onset of CAPS	2 nd day after foetal loss	4 th day after foetal loss
Organ involvement	Hepatic, Pulmonary, Placenta	Hepatic, Central nervous system, Placenta
Laboratory results at onsets of CAPS		
Haemoglobin (g/dL)	7.4	10.1
Platelets (x10⁹/L)	82	28
AST²/ALT³/LDH⁴ (U/L)	137/140/690	1199/1130/2206
Estimated proteinuria (g/day)	2.7	2.3
Positive serologies	Anti-Ro Anti-nuclear antibody, Anti-double stranded DNA, Lupus anticoagulant, Anti-cardiolipin IgM/IgG, B2 glycoprotein IgM/IgG	
Treatment	IV MP ⁵ , LMWH 1mg/kg/day twice daily	IV MP, PLEX, IVIG, LMWH 1mg/kg/day twice daily
Outcomes		
Obstetric	Intra-uterine foetal demise	
Maternal	Recovery	

¹Low molecular weight heparin. ²Aspartate aminotransferase. ³Alanine aminotransferase. ⁴Lactate dehydrogenase. ⁵Intravenous Methylprednisolone

Conclusion: Recognition of CAPS is complex in pregnancy. Vigilance is required for urgent aggressive treatment to improve outcomes in this uncommon condition.

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FRI0222 ANALYSIS OF CLINICAL AND SEROLOGICAL PICTURE OF PATIENTS WITH PRIMARY SJÖGREN'S SYNDROME AND AN EARLY DISEASE ONSET AT AGE BEFORE 35 YEARS

Qurania Argyropoulou¹, Andreas Goules¹, Evangelia Zampeli², Maria Mavromati³, Clio Mavragani^{1,4}, Fotini Skopouli^{3,5}, Haralampos M. Moutsopoulos^{1,2,6}, Athanasios Tzioufas^{1,2}. ¹National and Kapodistrian University of Athens, Pathophysiology, Athens, Greece; ²Institute of Autoimmune Systemic and Neurologic Diseases, Athens, Greece; ³Euroclinic Hospital, Internal Medicine and Clinical Immunology, Athens, Greece; ⁴National and Kapodistrian University of Athens, Physiology, Athens, Greece; ⁵Harokopio University of Athens, Athens, Greece; ⁶Academy of Athens, Athens, Greece

Background: Primary Sjögren's Syndrome (pSS) is a chronic systemic autoimmune disease with diverse clinical picture, extending from exocrinopathy to systemic disease and non-Hodgkin's lymphoma (NHL). Usually, it affects middle aged women, but early disease onset (≤ 35 years old) has been also observed. So far, there is a limited number of studies to explore whether the age of SS onset affects the clinical SS phenotype¹. To definitely answer this question a well-organized, large, harmonized cohort of patients with a continuous follow up is needed, through the ongoing European project (HarmonicSS, grant agreement no: 731944).

Objectives: To investigate whether the clinical and serological picture of pSS patients with early disease onset (≤ 35 years old) differs to that of middle-age onset.

Methods: Medical records of 717 pSS patients, included in two Greek cohorts fulfilling the 2002 revised European/American International classification criteria for SS, were evaluated. The study group included 133 (18.5%) patients with disease onset ≤ 35 years (median disease duration, range: 10, 0-32 years; group A) matched at 1:1 ratio according to gender and disease duration with 133 pSS patients with middle-age onset (mean

age at disease onset 52±5 years; median disease duration, range: 10, 0-30; group B). Clinical and laboratory data were collected through an extensive clinical chart review. All parameters were compared by chi-square or Fisher's exact test, when appropriate.

Results: The two pSS groups had similar frequencies of non-specific clinical findings (chronic fatigue, arthralgias-myalgias, arthritis, Raynaud's phenomenon), periepithelial manifestations (interstitial nephritis, lung and liver disease), splenomegaly, leucocyte and neutrophil counts. However, patients with disease onset ≤35 years old, exhibited increased proportion of salivary gland enlargement (SGE; 43,8% vs 25,4%, $p=0,003$), lymphadenopathy (18,2% vs 7,6%, $p=0,016$), palpable purpura (20,3% vs 9,8%, $p=0,025$), anti-Ro/SSA antibodies (90,2% vs 74%, $p=0,001$), rheumatoid factor positivity (73,3% vs 53,6% $p=0,002$), low C4 levels (64,5% vs 42,4%, $p=0,001$) and NHL (18% VS 8,3%, $p=0,028$).

Conclusion: Our findings suggest that young patients with pSS display increased prevalence of systemic features, B cell hyperreactivity, as well as heightened risk for lymphoma development. Further prospective studies with a larger number of patients are needed to address whether early disease onset may also serve as an additional risk factor for NHL development.

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FRI0223

EXTRACELLULAR VESICLES AS A SOURCE OF BIOMARKERS IN SJÖGREN'S SYNDROME: A SWATH-MS PROTEOMIC APPROACH

Chiara Baldini¹, Francesco Finamore², Francesco Ferro¹, Marta Mosca¹, Stefano Bombardieri¹, Silvia Rocchiccioli², Antonella Cecchetti¹. ¹University of Pisa, Department of Clinical and Experimental Medicine, Pisa, Italy; ²Institute of Clinical Physiology, CNR, Pisa, Italy

Background: Primary Sjögren's syndrome (pSS) is a multifactorial autoimmune disorder characterized by lymphocytic infiltration of the exocrine glands. In the recent past several proteomic studies attempted to look at valid biomarkers for pSS in whole saliva, however little is known about the composition of salivary extracellular vesicles (EVs), nor to what extent their content may reflect the phenotypic state of the disease.

Objectives: In this study, we search for pSS specific biomarkers by using a sequential window acquisition of all the theoretical fragment ion spectra (SWATH-MS) approach to monitor the dynamics of saliva EVs sub-proteome of pSS patients compared to healthy controls.

Methods: We included patients with a diagnosis of pSS made according to the AECG 2002 criteria and healthy volunteers as controls. Saliva was collected under standardized conditions. EVs were enriched by sequential ultracentrifugation steps from saliva samples. Peptide identification and quantitation was performed by matching SWATH data against an assay library of MS/MS spectra created from a previously acquired data-dependent acquisition (DDA) method. GO terms and protein-protein interaction network analysis were performed using Cytoscape software.

Results: We included 20 pSS patients (AECG 2002 criteria) and 10 healthy subjects. Quantitative data evidenced a distinct separation between the patient group and control group, indicating that pSS may modulate the phenotype of protein cargoes of salivary EVs. The majority of the proteins up-regulated in pSS compared to controls were found to be involved in several inflammatory processes. Particular emphasis was given to proteins converging to IL-12 signaling that included annexin A2, cofilin, macrophage migration inhibitory factor (MIF), S100A8-A9 and plasmin-2 proteins.

Conclusion: Our results revealed that the inflammatory phenotype observed in pSS patients is also extended to salivary EVs, which protein content may represent a novel source for potentially useful biomarkers for pSS.

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FRI0224

PREVALENCE OF SJÖGREN'S SYNDROME IN THE COMMUNITY OF MADRID

Jaime Barrio Cortes¹, Tomás Gómez Gascón¹, M.ª Dolores Esteban Vasallo², Isabel Del Cura González³, Juan Antonio López Rodríguez³, Ana Clara Zoni², María Felicitas Domínguez Berjón². ¹Fundación de Investigación e Innovación Biomédica de Atención Primaria, Madrid, Spain; ²Dirección General de Salud Pública, Madrid, Spain; ³Unidad de Apoyo a la Investigación de la Gerencia de Atención Primaria, Madrid, Spain

Background: Several studies on the epidemiology of autoimmune diseases performed within the past several decades revealed a prevalence of Sjögren Syndrome (SS) between 0.3% and 4.83% (1). More recent studies revealed a lower prevalence between 31/100,000 to 49/100,000 (2,3). The exact prevalence of SS is unknown due to the heterogeneity of the populations and geographic areas studied, the utilization of different diagnostic tests or the lack of a unique classification criteria for the disease. Also, the disease may have an insidious onset and a variable course with a broad spectrum of clinical manifestations, so the diagnosis may be delayed or SS patients may be missed and misclassified as other rheumatic disease. Besides, SS can occur alone (primary SS) or in association with other specific systemic autoimmune rheumatic diseases (secondary SS).

Objectives: The aim of our study was to determine the prevalence of SS in the Community of Madrid (CM) and to describe the sociodemographic and clinical characteristics of these patients.

Methods: Population-based cross-sectional study in the CM, Spain. The information source for SS cases was the Registry of rare diseases in the CM (SIERMA).

A descriptive analysis of the main sociodemographic and clinical characteristics of SS cases was performed.

Prevalence per 10,000 inhabitants in people with 18 years of age and over, stratified by sex and their 95% confidence intervals (CI) were calculated for 2015. The denominator was the people registered at the Population Register in the middle of the period (July 2015).

Results: There were 4,778 cases of SS in SIERMA and 389 (8,1%) of them were already dead. 4434 (92,8%) were women. The median age was 64,7 years (15,4), and the maximum age was 103 years. The disease was most frequent in the sixth decade with 1079 cases (22,6%), in the seventh decade with 1120 cases (23,4%) and in the eighth decade with 935 cases (19,6).

3116 cases (65,2%) were classified as primary SS (pSS) and 1662 (34,8%) as secondary SS (sSS).

Among the sSS the main rheumatological conditions associated were rheumatoid arthritis (58%), lupus (24,5%), systemic Sclerosis (10,5%); mixed connective tissue (4%), inflammatory muscle disease (2%) and vasculitis (1%).

The prevalence of SS in adults (≥ 18 years of age) was 8,4/10,000 with a 95% CI: 8,2 – 8,7 (14,9% in women and 0,8% in men). The prevalence of pSS was 5,5/10,000 (CI 5,3 – 5,7) and for sSS was 2,8/10,000 (95%CI: 2,7-3).

Conclusion: SS mainly affects females during the sixth, seventh and eight decades of life, and shows a female:male ratio of 9:1. Two out of three cases of SS identified were classified as pSS. The main rheumatological conditions associated were rheumatoid arthritis and lupus. The estimated prevalence of SS in the Community of Madrid population is lower than the observed in previous studies whereas the prevalence of pSS is similar than the observed in current studies.

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