Society of Nephrology/Renal Pathology Society. A delayed 48-hour gallium scan was performed and interpreted by visual and semiquantitative methods. The renal histological results were compared with gallium scan parameters.

Results: Out of 266 participants, 189 (71%) were proliferative LN. Baseline gallium left kidney/spine (K/S) ratio was significantly higher in class IV LN as compared to class III or class V (median [inter-quartile range, IQR]: 1.14 [1.0-1.3], 0.97 [0.9-1.1], 1.035 [0.9-1.2], respectively, p<0.001). Patients with higher daily urine protein, renal activity index, endocapillary proliferation and interstitial inflammation tended to exhibit higher gallium uptake by regression analysis. Besides, changes of gallium uptake ratio between 2 biopsies were positively correlated with daily urine protein change (r=0.699, p<0.001), change of activity index (r=0.339, p=0.033), endocapillary proliferation (r=0.380, p=0.027) and neutrophils infiltration (r=0.405, p=0.018) in renal pathology findings.

Table1. (N=266)

	Class I&II (n=7)	III (n=43)	IV (n=146)	V (n=70)	*P* value
Age	42.0 (33.0-43.0)	31.0 (26.0-40.0)	32.0 (25.0-41.0)	31.0 (26.0-40.3)	0.572
Gender					0.577
F	7 (100.0%)	36 (83.7%)	118 (80.8%)	59 (84.3%)	
M	0 (0.0%)	7 (16.3%)	28 (19.2%)	11 (15.7%)	
Lab data					
Daily urine protein	0.7 (0.1-1.8)	1.9 (1.0-2.5)	3.7 (2.1-5.6)	2.1 (0.9-6.3)	**<0.001****
Creatine	0.7 (0.5-0.8)	0.8 (0.6-1.2)	1.4 (0.9-2.8)	0.8 (0.7-1.1)	**<0.001****
eGFR	97.5 (91.0-139.2)	88.3 (65.6-125.6)	48.5 (22.3-79.0)	88.2 (60.8-117.6)	**<0.001****
dsDNAAb	248.7 (166.5-477.0)	169.0 (64.7-347.7)	265.4 (125.5-464.1)	126.0 (33.3-286.9)	**<0.001****
C3	78.2 (71.3-90.5)	76.4 (63.7-93.2)	54.6 (40.7-67.0)	67.9 (47.9-97.0)	**<0.001****
C4	15.1 (4.1-20.9)	14.4 (10.4-22.2)	10.6 (6.7-18.0)	12.5 (7.2-23.5)	**0.048***
SLEDAI	18.5 --	14.0 (10.0-14.0)	14.0 (8.0-20.0)	8.0 (5.5-10.8)	**0.025***
Renal pathology					
ActivityIndex	0 (0-1)	3 (1-4)	8 (5.0-10.3)	0 (0-2)	**<0.001****
Cellular crescent	0 (0-0)	0 (0-0)	2 (0-2)	0 (0-0)	**<0.001****
Necrosis/ Karyorrhexis	0 (0-0)	2 (0-2)	2 (0-2)	0 (0-0)	**<0.001****
Endocapillary proliferation	0 (0-0)	1 (0-1)	2 (2-3)	0 (0-1)	**<0.001****
Hyaline thrombus/wire loop	0 (0-0)	0 (0-0)	1 (0-2)	0 (0-0)	**<0.001****
Neutrophils infiltration	0 (0-0)	0 (0-0)	0 (0-1)	0 (0-0)	**<0.001****
Interstitital inflammation	0 (0-1)	0 (0-1)	1 (0-2)	0 (0-1)	**<0.001****
Chronicity Index	0 (0-1)	2 (0-3)	1 (0-4)	1 (0-2)	**0.036***
Glomerular stenosis	0 (0-0)	1 (0-1)	1 (0-2)	0 (0-1)	**0.001****
Tubular atrophy	0 (0-0)	1 (0-1)	0 (0-1)	0 (0-1)	0.094
Interstitial fibrosis	0 (0-1)	1 (0-1)	0 (0-1)	0 (0-1)	0.405
Fibrous crescent	0 (0-0)	0 (0-0)	0 (0-0)	0 (0-0)	0.066
Tubulointerstitial nephritis	1 (14.3%)	17 (39.5%)	83 (56.8%)	22 (31.9%)	**0.001****
Atherosclerosis	1 (14.3%)	9 (20.9%)	32 (21.9%)	8 (11.8%)	0.343
L't k/s ratio	0.9 (0.8-1.2)	1.0 (0.9-1.1)	1.1 (1.0-1.3)	1.0 (0.9-1.2)	**<0.001****
R't k/s ratio	1.0 (0.9-1.4)	1.0 (0.9-1.2)	1.2 (1.1-1.4)	1.1 (1.0-1.2)	**<0.001****
Gallium visual grading					**<0.001****
0-1&1&1-2	4 (57.1%)	19 (44.2%)	26 (17.8%)	18 (25.7%)	
2	1 (14.3%)	17 (39.5%)	42 (28.8%)	33 (47.1%)	
2-3	0 (0.0%)	6 (14.0%)	38 (26.0%)	10 (14.3%)	
3	2 (28.6%)	1 (2.3%)	40 (27.4%)	9 (12.9%)	

Chi-square test. †Kruskal-wallis test. *P <0.05, **P <0.01

Table2. LK ratio spine.(Multivariate1 Adjust R^2=0.219; Multivariate2 Adjust R^2=0.217; Multivariate3 Adjust R^2=0.225)

	Univariate			Multivariate1			Multivariate2			Multivariate3†		
	B	95%CI	*P* value	B	95%CI	*P* value	B	95%CI	*P* value	B	95%CI	*P* value
Age	-0.002	(-0.005, 0.000)	0.079									
Gender												
F	Refrence	Refrence										
M	0.004	(-0.076, 0.084)	0.925									
Lab data												
Daily urine protein	0.023	(0.016, 0.031)	**<0.001****	0.020	(0.012, 0.027)	**<0.001****	0.019	(0.011, 0.026)	**<0.001****	0.018	(0.011, 0.026)	**<0.001****
Creatine	0.016	(-0.002, 0.035)	0.088									
eGFR	-0.001	(-0.001, 0.000)	**0.016***	0.000	(-0.001, 0.001)	0.965	0.000	(-0.001, 0.001)	0.757			
dsDNAAb	0.000	(0.000, 0.000)	0.113									
C3	-0.001	(-0.002, 0.000)	**0.019***	0.001	(-0.001, 0.002)	0.370	0.001	(-0.001, 0.002)	0.257			
C4	-0.001	(-0.004, 0.001)	0.375									
SLEDAI	-0.005	(-0.020, 0.009)	0.448									
Renal pathology												
ActivityIndex	0.021	(0.015, 0.027)	**<0.001****	0.018	(0.010, 0.026)	**<0.001****						
Cellular crescent	0.045	(0.022, 0.068)	**<0.001****				0.000	(-0.029, 0.030)	0.995			
Necrosis/ Karyorrhexis	0.036	(0.017, 0.055)	**<0.001****				0.017	(-0.007, 0.040)	0.167			
Endocapillary proliferation	0.073	(0.048, 0.098)	**<0.001****				0.029	(-0.010, 0.069)	0.144	0.039	(0.011, 0.068)	**0.007****
Hyaline thrombus/wire loo	0.083	(0.049, 0.117)	**<0.001****				0.019	(-0.025, 0.064)	0.396			
Neutrophils infiltration	0.101	(0.053, 0.149)	**<0.001****				0.008	(-0.048, 0.065)	0.768			
Interstitital inflammation	0.091	(0.057, 0.125)	**<0.001****				0.051	(0.011, 0.091)	**0.013***	0.053	(0.016, 0.090)	**0.005****
Chronicity Index	-0.002	(-0.014, 0.009)	0.700									
Glomerular stenosis	-0.015	(-0.047, 0.017)	0.356									
Tubular atrophy	-0.010	(-0.045, 0.026)	0.592									
Interstitial fibrosis	-0.002	(-0.038, 0.034)	0.899									
Fibrous crescent	0.025	(-0.092, 0.143)	0.672									
Tubulointerstitial nephritis	0.020	(-0.041, 0.081)	0.519									
Atherosclerosis	-0.024	(-0.101, 0.054)	0.547									
Gallium visual grading												
0-1&1&1-2	Refrence	Refrence										
2	0.175	(0.134, 0.217)	**<0.001****									
2-3	0.343	(0.295, 0.390)	**<0.001****									
3	0.416	(0.568, 0.664)	**<0.001****									

Linear regression.†Stepwise. *P<0.05 **P<0.01

Conclusion: Higher renal gallium uptake predicts higher active inflammation in LN. Changes of gallium uptake ratio were correlated with changes histopathological parameters and daily urine protein. Renal gallium scan appeals to be a valuable non-invasive tool in assessing activities of LN.

REFERENCES

[1] Tsai, S. C., Hsieh, T. Y., Huang, P. W., & Lin, W. Y. (2016). Absolute quantitative evaluation of 67Ga scintigraphy in lupus nephritis. Clinical nuclear medicine, 41(6), 442-446.

[2] Lin, W. Y., Hsieh, J. F., Tsai, S. C., Lan, J. L., Cheng, K. Y., & Wang, S. J. (2000). Semi-quantitative evaluation of gallium-67 scintigraphy in lupus nephritis. European journal of nuclear medicine, 27(11), 1626-1631.

[3] Nomura, S., Watanabe, Y., Otsuka, N., & Osawa, G. (1996). Gallium 67 scintigraphy as a predictor of renal prognosis in primary immunoglobulin A nephropathy. American journal of kidney diseases, 27(2), 204-208.

Disclosure of Interests: Tsu-Yi Hsieh: None declared, Yi-Ming Chen Grant/research support from: GSK, Pfizer, BMS, Astra & Zeneca, Consultant for: Pfizer, Novartis, Abbvie, Johnson & Johnson, BMS, Roche, Sanofi, MSD, Guigai, Astellas, Inova Diagnostics, UCB, Agnitio Science Technology, United Biopharma, Thermo Fisher, Paid instructor for: Pfizer, Novartis, Abbvie, Johnson & Johnson, BMS, Roche, Astra& Zeneca, Sanofi, MSD, Guigai, Astellas, UCB, Thermo Fisher, Speakers bureau: Pfizer, Novartis, Abbvie, Johnson & Johnson, BMS, Roche, Lilly, GSK, Astra& Zeneca, Sanofi, MSD, Guigai, Astellas, UCB, Thermo Fisher, Wei-Ting Hung: None declared, Hsin-Hua Chen Speakers bureau: Johnson & Johnson, Novartis, Pfizer, Abbvie, Roche, UCB, Bristol-Myers Squibb, Chugai, Kuo-Lung Lai: None declared, Ching-Tsai Lin: None declared, Yi-Da Wu: None declared, Wen-Nan Huang: None declared, Chih-Wei Tseng: None declared, Yi-Hsing Chen: None declared
DOI: 10.1136/annrheumdis-2019-eular.8266

SAT0174 CHARACTERISTICS AND TREATMENTS OF PATIENTS WITH SJÖGREN'S SYNDROME IN A REAL-WORLD SETTING

Carlos Gonzalez-Najera[1], Steven Taylor[2], Sean Crawford[1], Premkumar Narasimhan[1], Xiao Shao[1], LI Jun[1]. [1]*Bristol-Myers Squibb, Princeton, United States of America*; [2]*Sjögren's Syndrome Foundation, Reston, United States of America*

Background: Sjögren's syndrome (SS) is one of the most common autoimmune diseases. Main symptoms include dry eyes and mouth, followed by difficulty swallowing and serious systemic manifestations including neurological, pulmonary and musculoskeletal effects.[1] However, SS is not homogeneous; it affects patients in different ways, with varying symptoms and severity. Identifying discrete patient types and understanding their commonalities and differences is integral to enhancing disease knowledge, identifying areas of unmet need, defining the size of the affected population and determining appropriate treatment approach.

Objectives: Using data gathered by the SS Foundation through a survey of over 3000 patients with SS, analyses were performed to: 1) describe patient characteristics and treatment in a real-world setting and 2) categorise patients based on baseline characteristics.

Methods: The analytical approach used clustering techniques to synthesise the mixed-data–type survey in two separate analyses. Data were filtered to include patients 40–65 years of age. Analyses were performed firstly to identify relationships between features—related to demographic, disease state, treatments, etc—and secondly to identify clusters of patients. A Bayesian network model was used to evaluate feature correlations and the probability of a specific survey answer given one or many preliminary conditions. With respect to patient clustering, the model used was based on Gower distance and Partitioning Around Medoids, which each can include both numerical and categorical data.

Results: We evaluated comorbidities, symptoms and treatment to identify traits that differed most significantly among patient types. Four discrete patient clusters were identified (disease severity measured by median EULAR SS Patient Reported Index score[2]; Table 1):

- 1. Recent: Most recently diagnosed, least severe disease manifestation
- 2. Slower progressing: Longest time since diagnosis, second–lowest disease severity
- 3. Second–second: Second in time since of diagnosis, second in disease severity
- 4. Most severe: Most severe disease manifestations and comorbidities

The most differentiating comorbidities across all clusters included gastroesophageal reflux disease, fibromyalgia and Raynaud's syndrome (Table 2). The symptoms that impacted patients' lives the most and that were differentiating across clusters were brain fog, fatigue and forgetfulness. The treatments that were most contrasting were oral comfort agents, followed by DMARDS and secretagogues (Figure 1). The patient cluster definitions above suggest some contradictions, because not all significant cluster characteristics progressively worsened with increasing disease severity.

Conclusion: These analyses show differences in disease characteristics and treatment across patient clusters; however, more work is required to validate these clusters.

REFERENCES

[1] Ienopoli S, et al. Oral and Maxillofac Surg Clin North Am 2014;26:91-9.

[2] Seror R, et al. Ann Rheum Dis 2011;70:968-72.

Table 1. Most significant patient features

	Recent	Slower progressing	Second-second	Most severe
Patients, n	154	243	184	132
Female, %	92	95	95	92
Age, years (median)	57	57	59	59
Years since diagnosis (median)	5	9	6	8
ESSPRI score (median)	4	5.3	6	6.5
Dryness (median)	6	6	7	7
Fatigue (median)	5	6	7	7
Pain (median)	2	4	5	6
Total hospitalisation (ER/UCC/hospital), %	14	17	22	27
HCP (median)	3	4	4	5
Total spending (mean $US)	1708	3020	3823	5550
Prescriptions, n (median)	2	2	3	5
OTC medications, n (median)	3	3	4	5
Awarded social security disability, %	2%	8%	9%	16%

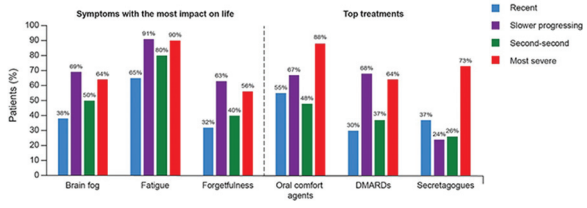
ER=emergency room; ESSPRI=EULAR SS Patient Reported Index; HCP=healthcare provider; OTC=over the counter; UCC=urgent care centre

Table 2. Top comorbidities that differentiate clusters

Comorbidity, %	Recent	Slower progressing	Second-second	Most severe
GERD	21	45	45	53
Pneumonia or bronchitis	8	12	10	18
Leucopoenia	8	16	11	16
Anaemia	11	17	10	17
Fibromyalgia	10	27	38	43
Hypertension	18	22	21	24
Sinusitis	14	31	38	49
Mixed connective tissue disease	4	13	12	14
Irritable bowel syndrome	14	32	33	35
Raynaud's disease	24	35	35	40

GERD=gastroesophageal reflux disease

Figure 1. Top three symptoms and treatments that differed most significantly among clusters



Acknowledgement: Professional medical writing: Fiona Boswell, PhD, Caudex; funding: Bristol-Myers Squibb

Disclosure of Interests: Carlos Gonzalez-Najera Shareholder of: Bristol-Myers Squibb, Employee of: Bristol-Myers Squibb, Steven Taylor: None declared, Sean Crawford Shareholder of: Bristol-Myers Squibb, Employee of: Bristol-Myers Squibb, Premkumar Narasimhan Employee of: Bristol-Myers Squibb, Xiao Shao Shareholder of: Bristol-Myers Squibb, Employee of: Bristol-Myers Squibb, Jun Li Shareholder of: Bristol-Myers Squibb, Employee of: Bristol-Myers Squibb

DOI: 10.1136/annrheumdis-2019-eular.1141

SAT0175 ULTRA-HIGH-FREQUENCY ULTRASOUND OF LABIAL SALIVARY GLANDS HIGHLY CORRELATES WITH HISTOPATHOLOGY IN PRIMARY SJÖGREN'S SYNDROME

Chiara Baldini[1], Francesco Ferro[1], Veronica Iodice[2], Rossana Izzetti[2], Valentina Donati[3], Silvia Fonzetti[1], Teresa Oranges[1], Valentina Dini[1], Dania Cioni[2], Nicoletta Luciano[1], Rosa Maria Bruno[1], Marta Mosca[1], Saverio Vitali[2], Davide Caramella[2]. [1]*University of Pisa, Department of Clinical and Experimental Medicine, Pisa, Italy*; [2]*University of Pisa, Radiology Unit, Pisa, Italy*; [3]*University of Pisa, Pathology Unit, Pisa, Italy*

Background: In the last few decades many studies have investigated the role of major salivary glands ultrasound (SGUS) in the diagnostic work-up of patients with suspected primary Sjögren's syndrome (pSS). Recent development of ultra-high-resolution ultrasound systems (UHFUS), with frequencies as high as 70 MHz and capability resolution as fine as 30 µm, has permit new diagnostic applications to a variety of superficial targets including labial salivary glands (LSGs). To date, however, no information are available regarding the use of UHFUS for the study of LSGs in humans.

Objectives: To evaluate the feasibility and the diagnostic accuracy of UHFUS of LSGs in patients with suspected SS and to assess the correlations between LSG histopathology, UHFUS and SGUS.

Methods: Consecutive patients with clinically suspected pSS were included in this study. All patients underwent a complete diagnostic work-up including conventional SGUS and LSG biopsy. The same expert pathologist calculated for all the samples the focus score (FS), number of foci and evaluated the presence of ectopic germinal centers (GCs). UHFUS of LSG was performed by specialized radiologists 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), fibrosis and eco color-Doppler vascular pattern and grade of vascular intensity.

Results: We included 32 patients with suspected pSS. At the end of the work-up, pSS diagnosis was confirmed in 12/32 (37.5%) cases. No differences between pSS patients and no-SS sicca controls were observed in UHFUS findings related to LSG distribution and eco color-Doppler vascular parameters. By contrast, pSS patients presented statistically significant differences in peripheral UHFUS inhomogeneity (p=0.006), and a higher degree of fibrosis (p=0.01). Considering a score $\geq$2 in parenchymal inhomogeneity as suggestive for pSS, UHFUS appeared less specific than conventional SGUS (UHFUS Sp=70% vs SGUS Sp=95%) but more sensitive (UHFUS Se=83.3% vs SGUS Se = 58.3%). In addition, when we investigated the relationship between UHFUS, SGUS and LSG histopathology we found that the correlation coefficients between UHFUS inhomogeneity and LSG FS (UHFUS r=0.706** *vs* SGUS r=0.393*), number of foci (UHFUS r=0.712** *vs* SGUS r=0.445*), and number of ectopic GCs (UHFUS r=0.525** *vs* SGUS r=0.141), were significantly higher than those observed with conventional SGUS.

Conclusion: the application of UHFUS to the study of LSG in pSS appeared feasible and sensitive. Because of the anatomical details obtained with this technique and its stronger correlation with LSG histopathology, UHFUS might offer unique advantages over the existing major salivary gland imaging modalities in pSS assessment.

Disclosure of Interests: Chiara Baldini: None declared, Francesco Ferro: None declared, Veronica Iodice: None declared, Rossana Izzetti: None declared, Valentina Donati: None declared, Silvia Fonzetti: None declared, Teresa Oranges: None declared, Valentina Dini: None declared, Dania Cioni: None declared, Nicoletta Luciano: None declared, Rosa Maria Bruno: None declared, Marta Mosca Paid instructor for: GlaxoSmithKline, Lilly, UCB, Saverio Vitali: None declared, Davide Caramella: None declared

DOI: 10.1136/annrheumdis-2019-eular.6157

SAT0176 MACROPHAGE ACTIVATION SYNDROME IN MONOGENIC LUPUS: A RARE COMPLICATION OF A RARE DISEASE

Serkan Turkucar[1], Tugce Tunca Kucukali[2], Asena Baserdem[2], Erbil Unsal[1]. [1]*Dokuz Eylul University, Pediatric Rheumatology, Izmir, Turkey*; [2]*Dokuz Eylul University, Faculty of Medicine, Pediatrics, Izmir, Turkey*

Background: Macrophage activation syndrome (MAS) is one of the most severe, potentially life-threatening complication of childhood rheumatic diseases, especially SoJIA and J-SLE. MAS in the course of J-SLE has a high mortality rate (8%-22%) [1]; however, it is difficult to differentiate from the SLE flare itself.