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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
Leucopenia	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

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SAT0175

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

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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 ≥ 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.

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SAT0176

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

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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.