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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%

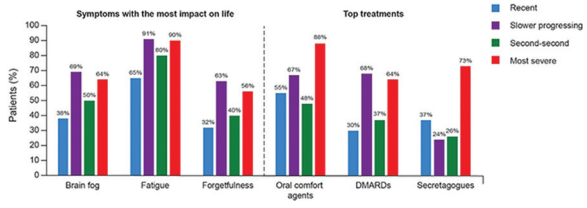
ER=emergency room; ESSPRI=EULAR SS Patient Reported Index; HCP=healthcare provider; OTC=over the counter; UCC=urgent care centre

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
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



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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 $\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.

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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.

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Objectives: This case report is presented to emphasize that the deterioration of certain clinical and laboratory findings of an SLE patient should alert the physician for development of MAS.
Methods: We reported a monogenic lupus patient with MAS clinic with acute pancreatitis, who had c1qA gene mutation.
Results: A 9-year-old male was first referred with photosensitivity and erythematous crusted lesions on his extremities and face while he was 2 years old. ANA was positive (1/1000 dilution) and skin biopsy from lesions was compatible with subacute lupus. Steroid, metotrexate and hydroxychloroquine were administrated. Patient's male identity and atypical age for lupus was further evaluated with "whole exome sequencing" method. Complement c1qA gene mutation was detected. Monthly fresh frozen plasma and IVIG treatments were added, which able to control the disease to some extent.
He had acute gastroenteritis and fever, leading to activation of skin and oral mucosa lesions with on bilateral parotitis in his last visit (Figure-A). His body temperature was 39.9 °C. USG confirmed the parotitis. Regarding infectious gastroenteritis, IVIG and plasma with broad-spectrum antibiotics were administrated. Despite treatment, high fever persisted. He had hypotension, irritability, and hepatosplenomegaly with abdominal tenderness on physical examination. Abdomen CT was compatible with acute pancreatitis (Figure-B), and he had high amylase and lipase levels. Despite IV hydration with intense therapy, developing pancytopenia, hypoalbuminemia, increase in hepatic enzymes, decrease in fibrinogen and ESR with elevated CRP and ferritin levels were alerts for MAS. Bone marrow aspiration demonstrated hemophagocytosis (Figure-C). Pulse high dose IV methyl prednisolone treatment was started. Clinical and laboratory findings, particularly ferritin level, improved in the third day of treatment. Steroids were gradually reduced while cyclosporine was added. He is under remission with low dose steroid, cyclosporine-A and regular plasma infusions per 3 weeks.

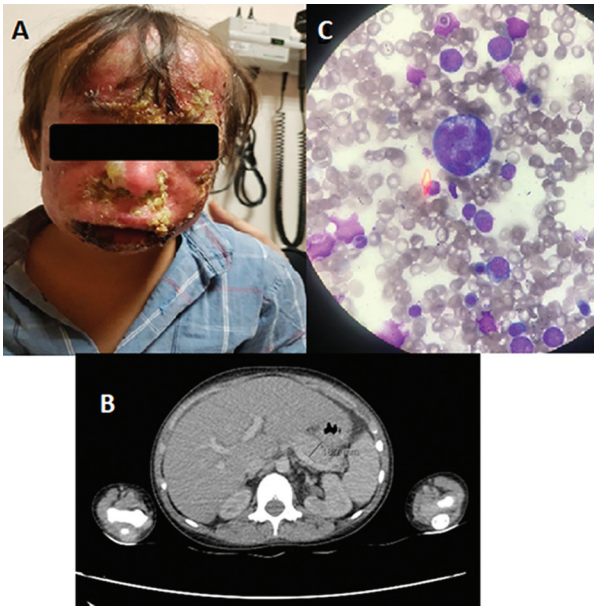


Figure. **A**. Bullous and crusted lesions on face and around oral mucosa. **B.** Shortening of the pancreas tail in CT. **C.** Hemophagocytic cells in the bone marrow.

Conclusion: MAS is rarely seen in course of monogenic J-SLE, but may be serious. It is the first juvenile monogenic lupus case with MAS in the English literature, to best of our knowledge. It is important to distinguish this mortal clinical condition from the flare of disease and appropriate treatment should be started as soon as possible.

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SAT0177

CLINICAL AND EPIDEMIOLOGICAL RELEVANCE OF ALLERGY IN SYSTEMIC LUPUS ERYTHEMATOSUS: AN OBSERVATIONAL STUDY

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Background: Altered innate and adaptive immune response is a hallmark of the pathogenesis of systemic lupus erythematosus (SLE). Autoimmune manifestations are a major cause of morbidity and damage accrual and constitute the main target of treatment in patients with SLE. Exposure to medications for purposes of immune suppression or treatment of complications might also synergise with an aberrant immune response and facilitate infections as well as hypersensitivity to non-harmful antigens. Epidemiological data about allergy in SLE are scanty.
Objectives: To describe clinical and epidemiological features of allergy and asthma in SLE.
Methods: One-hundred-thirty-five patients with SLE (classified according to the American College of Rheumatology, ACR, 1990 revised criteria) were enrolled. Retrospective data collection included autoimmune manifestations during the course of SLE and history of asthma. In addition, we recorded timing, clinical features, culprit(s) of the first three hypersensitivity reactions (HyR, together with concomitant infections, SLEDAI-2K and prednisone dose at time of each reaction. Damage accrual was quantitated by employing the SLE International Collaborating Clinics/ACR damage index (SDI).
Results: A history of one or more HyR was found in 40.7% of patients. Immediate-type HyR were most significantly represented (83.6% of cases) and four cases were anaphylaxis (lifelong incidence rate of 67.04/105 persons-years). Respiratory allergy (allergic rhinitis or asthma) occurred in 19 patients (34.5%). Twenty-one out of 55 (38.3%) patients with allergy had $\geq$2 manifestations, five (9.1%) had $\geq$3 manifestations. Drugs were the most frequent culprits, accounting for 81.8% of HyR. Early-onset SLE (before the age of 21) associated with a higher risk of becoming allergic during SLE course compared with late-onset SLE (LR=8.01; p=0.005; HR=2.28, 95%CI=1.27-4.11). Patients with allergy onset after SLE (52.7% of cases) developed a second HyR more frequently and earlier (LR=11.49; p=0.001; HR=5.19, 95%CI=1.87-14.42). SLE with vasculitic manifestations also was a risk factor for developing HyR (LR=4.00; p=0.045; HR=2.13, 95%CI=1.00-4.55). Patients with allergy post-lupus had a SLEDAI-2K>4 and a prednisone dose>5mg/day in 51.7%, 41.7%, 50% of cases and in 37.9%, 50% and 50% of cases at first, second and third HyR respectively. Concomitant infections were detected in 20.7, 25% and 0% of cases respectively. Patients with $\geq$2 HyR had faster damage accrual compared to patients with $\leq$1 manifestations (median SDI/year=0.04, IQR=0.00-0.11 vs 0.00, IQR=0.00-0.06; p = 0.048).
Conclusion: Patients with SLE have a high prevalence of allergy, which might contribute to its protean phenotype SLE deflects allergy incidence trajectories towards an increased risk. Allergy, in turn, associates with increased SLE activity, medication burden and infectious complications and might affect the risk of long-term damage accrual.

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