Conclusions: Our results demonstrated that a clinically and persistent CAD triples the risk of damage compared to milder or relapsing courses, while hydroxychloroquine appeared to have a "protective" effect. Identifying the prevailing pattern of disease activity in every patient can be translated into a more effective personalized preventing strategy to reduce damage accrual and improve outcomes.

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FRI0256 ULTRASOUND CONSENSUS DEFINITIONS ON NORMAL AND ABNORMAL FINDINGS IN SALIVARY GLANDS IN SJÖGREN'S SYNDROME: RESULTS OF AN OMERACT DELPHI PROCESS

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Background: Ultrasonography (US) of salivary glands (USSG) may support diagnosis and evaluation of disease activity in primary Sjögren's syndrome pSS. In order to extent the use of USSG in daily practice, consensus definitions of normal and abnormal USSG findings are needed.

Objectives: To obtain consensual definitions of normal and abnormal USSG findings using a Delphi process.

Methods: Twenty-seven experts participated in the Delphi process. A Likert scale from 1–5 was used in which 1 indicated complete disagreement and 5 complete agreement. Three core items were proposed: 1. Consensus of assessment of SG (parotid gland (PG), submandibular glands (SMG), sublingual glands (SLG)). 2. Consensus for elementary lesions of PG, SMG, and SLG changes in PSS patients. 3. Consensus on grading of PG, SM, and SLG in pSS.

Results: The consensus of definitions between experts was reached after three rounds. The results (table 1) showed agreement with more than 75% of agreement in normal and abnormal definitions in SG and concluded to not evaluate SLG in pSS patients as well as to score pSS SG patients with at least two abnormal glands out of 4 (2 PG and 2 SMG) with a scoring system >2 for each abnormal gland.

Conclusions: We developed a consensual US definition of normal and abnormal USGS findings through a Delphi exercise process. This finding will be used as a basis to further proceed with the validation of USGS for clinical application.

Disclosure of Interest: None declared

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FRI0257 URINARY ANGIOSTATIN, CXCL4 AND VCAM-1 AS BIOMARKERS FOR LUPUS NEPHRITIS

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Abstract FRI0256 – Table 1

(% of agreement)	Definition of normal US findings	Procedure of scanning	Definition of abnormal US findings	SG to evaluate in pSS	Definition of scoring
Parotid glands	Uniformly echoic texture with a clear delineation from the superficial tissue. Tissue comparable to thyroid parenchyma (81%)	Longitudinal and transverse plane (90%)	Focal or diffuse an/hypoechoic areas (95%)	yes	4-grade semiquantitative scoring system (i.e. grade 0, normal parenchyma; grade 1, minimal change; grade 2, moderate; grade 3, severe; diffuse inhomogeneity occupying all the surface of the gland) (79%)
Submandibular glands	SMG are usually of finer granular echo texture compared to PG or to the normal thyroid parenchyma (89.1%)	Longitudinal and transverse plane (90%)	Idem PG (95%)	yes	Idem PG (79%)
Sublingual glands	SLG has no clear delineation from the superficial tissue because of no true fascial capsule (77%)	longitudinal and transverse plane (77%)	Idem PG (95%)	no	Not useful

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Background: Our preliminary proteomic analysis revealed elevated levels of urinary angiotensin, CXCL4 and VCAM-1 in patients with active lupus nephritis

Objectives: To study the performance of urinary angiotensin, CXCL4 and VCAM-1 in differentiating between active renal and non-renal SLE.

Methods: Patients who fulfilled the ACR classification for SLE with active renal disease, active non-renal disease and inactive disease were randomly recruited, together with a group of healthy controls. Stored urine samples of the participants were assayed for angiotensin, CXCL4 and VCAM-1 and compared among the different groups. ROC analysis was performed to obtain the best cut-off values to calculate the performance of these markers in differentiating among the different groups of patients. SLE disease activity was assessed by the SELENA-SLEDAI and physician's global assessment (PGA). Specificity and sensitivity of these markers in differentiating between active renal and non-renal SLE was determined by 2x2 contingency tables. In patients with active renal disease, correlation between these urinary biomarkers with clinical renal parameters was also performed.

Results: 227 SLE patients (80 inactive SLE; 67 active non-renal disease; 80 active renal disease; 94% women, age 39.2±13.8 years) and 53 controls were studied. Urinary angiotensin, CXCL4 and VCAM-1 levels normalized for creatinine were significantly higher in patients with active renal than non-renal disease (angiotensin 18.4±27.1 vs 1.6±2.91 pg/ng; p<0.0001; CXCL 9.11±15.7 vs 5.12±10.4 pg/ng; p=0.003; VCAM-1.41±1.31x10³ vs 0.72±1.10x10³ pg/ng; p<0.0001). The levels of these urinary protein markers were also significantly higher in active SLE patients than inactive SLE patients or healthy controls. Urinary angiotensin, CXCL4 and VCAM-1 correlated significantly with the renal SLEDAI (Rho 0.66, 0.45 and 0.51, respectively; p<0.001 in all), total SLEDAI score (Rho 0.60, 0.46 and 0.53, respectively; p<0.001 in all) and urine protein-to-creatinine (uPCR) ratio (Rho 0.73, 0.51, 0.59, respectively; p<0.001) in all the SLE patients studied. Urinary angiotensin was more specific (specificity 0.82) than elevated anti-dsDNA and low C3 (specificity 0.64 and 0.66) in differentiating active renal SLE from non-renal SLE. In a subset of patients with biopsy proven active lupus nephritis (N=68), these urinary protein markers could not differentiate between proliferative (III/IV) from non-proliferative (I/II/V) types of lupus nephritis. However, urinary CXCL4 (Rho 0.25; p=0.049) and VCAM-1 (Rho -0.28; p=0.02), but not angiotensin (Rho 0.11; p=0.39), correlated significantly with the histologic activity index. There was no significant association between these protein markers and the histologic chronicity index or renal SLEDAI score. On the other hand, urine angiotensin levels (Rho 0.36; p=0.003), but not CXCL4 (Rho 0.07; p=0.59) or VCAM-1 (Rho -0.11; p=0.36), correlated significantly with the uPCR ratio in this subgroup of patients.

Conclusions: Urinary angiotensin, CXCL4 and VCAM-1 are potentially useful biomarkers for SLE, in particular lupus nephritis. Further longitudinal studies are necessary to delineate the sensitivity and specificity of these two urinary protein markers in predicting renal flares and prognosis in SLE patients.

Disclosure of Interest: None declared

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FRI0258 LONG-TERM PROGNOSIS OF PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS-ASSOCIATED PULMONARY ARTERIAL HYPERTENSION: CSTAR-PAH COHORT STUDY

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Background: SLE-associated pulmonary arterial hypertension (PAH) is common in Asian countries, and the clinical outcome of patients with SLE-associated PAH is dramatically impaired.

Objectives: This study aimed to identify the long-term clinical outcomes and prognostic factors of patients with SLE-associated PAH confirmed by right heart catheterization (RHC).

Methods: A multicenter cohort of SLE-associated PAH was established. Baseline and follow-up records were collected. The primary endpoint was death from any

cause. The secondary experimental end point was treatment goal achievement (TGA), defined as an integrated outcome. **Results:** Among the 310 patients enrolled from 14 PAH centers, the median follow-up was 24.0 months. The 1-, 3- and 5-year survival rates were 92.1%, 84.8% and 72.9%, respectively. The 1-, 3- and 5-year TGA rates were 31.5%, 53.6% and 62.7%, respectively. Serositis, 6MWD >380 m and CI ≥ 2.5 L/min $\times$ m² were identified as independent prognostic factors of TGA. TGA within 5 years was identified as a factor associated with survival in patients with SLE-associated PAH (Figure 1).

Table 1. Baseline characteristics of patients with SLE-associated PAH

Characteristics	SLE-associated PAH (n=310)
Age at recruitment, yr	35.0 $\pm$ 10.1
Female sex, %	99.4
WHO Fc I-II, %	51.7
6MWD, m	408.6 $\pm$ 98.0
NT-proBNP, pg/ml	1660.5 $\pm$ 2275.1
SLEDAI	6.1 $\pm$ 5.5
mPAP, mmHg	46.5 $\pm$ 12.1
CI, L/min $\times$ m ²	2.8 $\pm$ 0.9
RAP, mmHg	5.8 $\pm$ 5.6

Conclusions: TGA was associated with the long-term survival, which supports and provides evidence to the treat-to-target strategy in SLE-associated PAH. Early diagnosis, intervention and heart function preservation are priorities for better long-term outcomes.

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FRI0259 LUPUS LOW DISEASE ACTIVITY STATE (LLDAS) IN PATIENTS WITH LUPUS NEPHRITIS (LN)

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Background: Lupus Low Disease Activity State (LLDAS) is a consensus-based definition of minimally acceptable disease activity in patients with Systemic lupus erythematosus (SLE).

Objectives: The aims of this study were to evaluate what proportion of patients with LN fulfils the definition of LLDAS and to evaluate the effect of LLDAS attainment on damage accrual over a period of 5 years.

Methods: This is a retrospective analysis of data prospectively collected in a longitudinal observational cohort of LN patients. The conceptual definition of LLDAS is a state, which if sustained, is associated with good long-term outcomes. The operational definition of LLDAS is fulfilled when all of the following criteria are met: (1) SLE Disease Activity Index (SLEDAI-2 K) ≤ 4 , with no activity in major organ systems (renal, central nervous system, cardiopulmonary, vasculitis, fever) and no haemolytic anaemia or gastrointestinal activity; (2) no new features of lupus disease activity compared to the previous assessment; (3) a Safety of Estrogens in Lupus Erythematosus National Assessment (SELENA)-SLEDAI physician global assessment (PGA) (scale 0–3) ≤ 1 ; (4) a current prednisolone (or equivalent) dose ≤ 7.5 mg daily; and (5) well-tolerated standard maintenance doses of immunosuppressive drugs and approved biologic agents, excluding investigational drugs. The definition of LLDAS was applied to each patient for each visit; organ damage was calculated with the SLICC/DI score (SDI) at study entry and at last observation.

Results: 294 patients were eligible for the study (89.79% females, mean age at

first visit 31.4 $\pm$ 11.9 years, mean disease duration at last visit 19.8 $\pm$ 8.7 years). At last observation 219 patients (85.37%) were on treatment with glucocorticoids (GC) and/or immunomodulators. According with all the items of the definition, at last observation LLDAS was present in 146 patients (49.65%); among these, 37 patients (20.8%) maintained a stable LLDAS during the last 5 years of follow-up (LLDAS fulfilled for all visits). 68 patients (23.12%) accrued organ damage during the follow-up; in the cohort as a whole the mean increase in SDI was 0.4 $\pm$ 0.67 resulting in a mean final SDI of 1.2 $\pm$ 1.8. Patients who maintained LLDAS were younger ($p < 0.05$), had a lower disease activity score at study entry ($p < 0.001$) and were more likely GC-free at last observation ($p < 0.001$). Patients who maintained LLDAS accrued less organ damage but this difference did not reach statistical significance.

Conclusions: High percentage of patients fulfils the proposed definitions for LLDAS at last visit but only a minority maintained this state for all the follow-up period. A minimally acceptable disease activity state is associated with a successful GC tapering and, probably, better long-term outcomes. LLDAS was shown to be associated with protection from damage accrual.

Disclosure of Interest: None declared

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FRI0260 ULTRASOUND STUDY OF PLEURAL PROFILE AND CHEST HIGH-RESOLUTION COMPUTED TOMOGRAPHY (HRCT): DIAGNOSTIC ROLE IN PRIMARY SJÖGREN'S SYNDROME-INDUCED LUNG INVOLVEMENT

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Background: Ultrasound pleural irregularity (PI-US) is a novel promising tool for non-invasive diagnosis of interstitial lung involvement (ILD) in connective tissue diseases (CTDs). Few data are available on its diagnostic usefulness in primary Sjögren's syndrome (pSS)-induced ILD.

Objectives: a) To assess the accuracy of PI-US to diagnose ILD in pSS when compared to chest tomography (HRCT) (i.e. gold standard imaging technique); b) to explore PI-US diagnostic value in early preclinical phases of lung involvement.

Methods: PI-US was performed by a single operator using a MyLab-25 (Esaote), 10 MHz, 5 cm linear probe. PI was defined as the loss of the normal hyperechoic linear pleural contour (score 0–2: normal, minimal and major changes at each intercostal space). Abnormal findings at HRCT were quantified by an expert radiologist according to a semiquantitative score (0–2: absent, moderate, severe). Semi-quantitative scores assigned by PI-US and HRCT to 6 lung fields (2 for the anterior, 2 for postero-superior and 2 for postero-inferior chest surface) were compared. Total and partial scores (for each lung fields) were evaluated. For statistical analysis chi-square, Mann-Whitney test, R-Spearman, and ROC-curve analysis were used.

Results: *Validation study phase (PI-US vs HRCT):* To validate PI-US technique, we enrolled 32 pSS patients [M/F:5/27; median age (IQR): 67 yrs (51.5–71); median disease duration (IQR): 7 (4–11) yrs; anti-Ro/SSA (+) 78.1%]. To be included patients should have performed a HRCT evaluation within 6 months. Thirteen patients (41%) presented HRCT lesions suggestive for ILD. HRCT total scores and PI-US total scores were strongly correlated ($r=0.744$, $p=0.000$). Similarly, PI-US and HRCT partial scores related to the postero-inferior fields showed a strong correlation one to each other ($r=0.780$, $p=0.000$). ROC-curve analysis identified a total PI-US score of 28.5 (Youden index) as able to predict HRCT-diagnosis of ILD with a sensitivity (SE) of 84.6% and a specificity (SP) of 89.5%. Analogously, a postero-inferior PI-US score of 12.5 demonstrated a SE of 100% and a SP of 89.5% for the ILD diagnosis. *Prospective study phase exploring the usefulness of PI-US for ILD early pre-clinical diagnosis:* We included 24 consecutive pSS patients without overt respiratory symptoms [M/F:1/23; median age (IQR): 55 yrs (47–67); median disease duration (IQR): 4 yrs (1–12); anti-Ro/SSA (+) 60.9%]. Out of them, at the end of the diagnostic work-up, four new cases of HRCT-proven pSS-ILD were diagnosed. Their PI-US mean total score was significantly higher than that observed in non-ILD patients (48 $\pm$ 18 vs 16 $\pm$ 12, $p=0.001$) as well as their mean postero-inferior PI-US score (19 $\pm$ 9 vs 6 $\pm$ 5, $p=0.003$). The total PI-US and postero-inferior PI-US cut-off retrieved in the first part of the study (i.e. 28.5 and 12.5) allowed us to identify those patients with an HRCT-proven pSS-ILD with a SE of 75% and 100%, a SP of 95% and 89.5%, a PPV of 75% and 57% and a NPV of 95% and 100%, respectively.

Conclusions: Even if preliminary, this study demonstrated a strong correlation between PI-US and HRCT in the detection of ILD-pSS also in asymptomatic patients, opening new perspectives for the early non-invasive screening of lung involvement in pSS.

Disclosure of Interest: None declared

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