Methods: Data were derived from electronic health records for all SLE patients seen in a university-based rheumatology clinic between 2013 and 2018. We evaluated the PF10a's floor and ceiling effects among different racial/ethnic groups. Construct validity was assessed by examining Spearman's correlation coefficients between the PF10a and other patient-reported (pain (scale 0-10) and pain visual analogue scale (VAS) (scale 0-100)), physician-reported (SLE disease activity index (SLEDAI)) and laboratory (erythrocyte sedimentation rate (ESR)) measures. Known-group validity was assessed by evaluating effect size (Cohen's d) between categories of pain (no pain vs. moderate-severe pain). We used standardized response means to examine the responsiveness of the PF10a to longitudinal changes in pain and SLEDAI. MID was estimated using distribution based and anchor-based methods.

Results: We included 612 patients in cross-sectional analyses of validity and 462 patients in longitudinal analyses of responsiveness. Mean age was 40.5±14.6, 87% were female and 32% Caucasian. The PF10a had ceiling effects above the commonly accepted criteria of 15% among Caucasian (23%), Asian (23%) and Other (17%) race/ethnicities, and no floor effects. Construct validity analyses showed strong correlations ($\rho=0.66$, $p<0.05$) with pain VAS, moderate correlations ($\rho=0.58$, $p<0.05$) with pain, and weak correlations with ESR ($\rho=0.25$, $p<0.05$) and SLEDAI ($\rho=0.16$, NS). Known-group validity analyses showed large differences among pain groups (Cohen's $d=1.49$, $p<0.05$). The PF10a was responsive to improvements in pain (SRM=0.5) and SLEDAI (0.49), but less so to deteriorations in pain (SRM=-0.42) or SLEDAI (SRM=-0.24). Distribution-based MIDs were +8 for improvement and -7 for deterioration. Anchor-based MIDs were +2 for improvement, -3 for deterioration with pain as anchor and +5 for improvement, -5 for deterioration with SLEDAI as anchor.

Conclusion: Although the PF10a showed some ceiling effects, it had good validity in this young racially/ethnically diverse sample with SLE. The PF10a was responsive to improvements in pain and disease activity. The anchor-based MIDs appear to be similar to those reported for PF10a in rheumatoid arthritis(1). This information supports the use of the PF10a in SLE and provides important information to facilitate interpretation of scores.

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SAT0195 SYSTEMIC DISEASES ASSOCIATED WITH AUTOIMMUNE LIVER DISEASES

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Background: Autoimmune liver diseases are considered as a group of hepatobiliary injuries mediated by abnormal immunity in subjects with genetic predisposition. It mainly includes autoimmune hepatitis, primary biliary cholangitis, primary sclerosing cholangitis and overlap syndrome. They are oftently associated with different manifestations of autoimmune systemic diseases such as systemic lupus erythematosus, CREST syndrome

Objectives: The aim of this study was to evaluate the prevalence and the characteristics of systemic diseases associated with autoimmune hepatopathies

Methods: We carried out a retrospective study from January 1996 to December 2018 including all patients with autoimmune liver diseases followed in our department: 64 primary biliary cholangitis (PBC), 22 autoimmune hepatitis (AIH), 23 overlap syndrome (OS), 15 primary sclerosing cholangitis (PSC).

Results: One hundred and twenty four patients were included in our study : 107 women and 17 men with an average age of 55 years [18-87 years]. An immunological disorder was found in 58 cases (48%): [28 PBC/12 AIH/12 OS/6 PSC] with a female predominance (Sex ratio (Men/

Women) = 0.09). Systemic diseases included respectively: Sjögren's syndrome in 20 cases, systemic lupus erythematosus in 7 cases, rheumatoid arthritis in 5 cases, CREST syndrome in 11 cases and autoimmune thyroiditis in 2 cases. The discovery of autoimmune disorder was in most cases concomitant to the diagnosis of the autoimmune liver disease (67%). Our study showed biological assessment perturbation in 20 cases with positivity of : anti-centromere antibodies (11 cases), anti-SSA antibodies (5 cases), anti-cardiolipin antibodies (4 cases) and rheumatoid factor (5 cases)

Conclusion: Our study shows a strong prevalence of systemic disease during autoimmune hepatopathies. A systematic screening for clinical and biological immune manifestations are highly recommended in those patients.

Disclosure of Interests: None declared

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SAT0196 RENAL FLARE IN CLASS V LUPUS NEPHRITIS: INCREASED RISK IN PATIENTS WITH TUBULOINTERSTITIAL LESIONS

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Background: Lupus nephritis leads to considerable morbidity and mortality in patients with systemic lupus erythematosus. Although membranous lupus nephritis has a better renal prognosis than proliferative lupus nephritis, progression to end-stage renal disease does occur. Renal flare is an independent predictor of renal prognosis in membranous lupus nephritis.

Objectives: To investigate the risk factors of renal flare in patients with membranous lupus nephritis.

Methods: Biopsy-proven pure membranous lupus nephritis patients diagnosed between January 1997 and September 2017 were studied. We assessed and compared the clinical and pathological parameters between patients who experienced renal flare and those who did not. To identify risk factors of renal flare, multivariable Cox proportional hazard regression analysis with stepwise backward elimination was performed.

Results: Out of the 53 patients with pure membranous lupus nephritis, 17 patients (32.1%) experienced renal flare during a median follow-up of 121.5 months (range, 44.4 – 196.9). Patients who experienced renal flare had significantly higher proportion of tubulointerstitial inflammation at baseline (76.5% vs. 36.1%, $p = 0.006$) and tubular atrophy/interstitial fibrosis (70.6% vs. 27.8%, $p = 0.003$). In multivariable Cox proportional hazard regression analysis, presence of tubulointerstitial inflammation (adjusted hazard ratio (HR) 5.532, 95% confidence interval (CI) 1.722 – 17.776, $p = 0.004$) and tubular atrophy/interstitial fibrosis (adjusted HR 4.328, 95% CI 1.450 – 12.916, $p = 0.009$) at baseline were significantly associated with increased risk of renal flare.

Conclusion: Presence of tubulointerstitial inflammation and tubular atrophy/interstitial fibrosis is associated with increased risk of renal flare in patients with membranous lupus nephritis. Closer monitoring of membranous lupus nephritis patients with tubulointerstitial lesions could lead to earlier treatments and better outcomes.

Table. Multivariable Cox proportional hazard regression analysis evaluating risk factors of renal flare in membranous lupus nephritis

	Adjusted HR	95% CI	P value
Urine WBC $\geq$ 3-5/HPF	0.854	0.193 – 3.774	0.835
Lupus anticoagulant	1.118	0.039 – 4.040	0.865
Activity index (glomerular component)†	1.218	0.621 – 2.387	0.567
Chronicity index (glomerular component)*	1.135	0.752 – 1.712	0.547
Tubulointerstitial inflammation	5.532	1.722 – 17.776	0.004
Tubular atrophy/Interstitial fibrosis	4.328	1.450 – 12.916	0.009

Variables with P value less than 0.1 in the univariable analysis were selected for inclusion in the multivariable analysis.

†Glomerular component of activity index, rather than total activity index, to avoid multicollinearity with tubulointerstitial inflammation

*Glomerular component of chronicity index, rather than total chronicity index, to avoid multicollinearity with tubular atrophy/interstitial fibrosis

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SAT0197 **INCIDENCE AND OUTCOME OF NON-TYPHOID SALMONELLA BACTEREMIA IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS IN COMPARISON WITH RHEUMATOID ARTHRITIS AND PRIMARY SJÖGREN'S SYNDROME**

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Background: Non-typhoid Salmonella (NTS) infection is an important problem in patients with systemic lupus erythematosus (SLE) [1, 2]. However, less is known about the incidence rate (IR) for NTS bacteremia.

Objectives: To evaluate the IR of NTS bacteremia in patients with SLE, in comparison to rheumatoid arthritis (RA) and primary Sjögren's syndrome (SjS), and by comparison with other Gram-negative bacilli (GNB) bacteremia including *Escherichia coli* (E. coli) and *Pseudomonas aeruginosa* (PsA). Furthermore, to access the survival outcome after NTS bacteremia.

Methods: In this single-center retrospective cohort study, we analyzed the registry of catastrophic illnesses database for patients fulfilled the criteria of SLE, RA, and SjS in National Taiwan University Hospital. The patients aged over 20 years, with adequate follow-up during January 1, 2008 to December 31, 2017 were included. Only new episodes of positive blood cultures during the observation period were taken into analysis. Cox proportional hazards regression model was applied for calculating the incidence rate ratio (IRR) of bacteremia between different diseases, and the hazard ratio (HR) of mortality.

Results: This study included 5953 patients. NTS bacteremia happened in 33 cases, among which 27 (81.8%) were group D1 and 4 (12.1%) were group B Salmonella. The baseline characteristics, IR of each GNB bacteremia, and the IRR adjusted for age, sex, diabetes mellitus (DM) as well as chronic kidney disease (CKD) are shown in Table 1. Compared to patients with RA, the IRR for NTS bacteremia was highest in SLE. The IRR for E. coli bacteremia in patients with SLE also increased in comparison with RA, although not as high as for NTS bacteremia. When compared to SjS, patients with SLE still had an elevated IRR of 3.52 (95% CI: 1.29–9.63, $p=0.014$) for NTS bacteremia. In SLE patients, DM increased the IR for NTS bacteremia (IRR: 4.43, 95% CI: 1.71–11.43, $p=0.002$; adjusted for age and sex).

In the 33 cases with NTS bacteremia [male: 21.2%; age: 49.0 (37.9–64.2) years], the median observation time to event was 11.1 (IQR, 3.0–38.1) months. No recurrent bacteremia was found. There were 14 (42.4%) deaths, and 7 (21.2%) died within 60 days after NTS bacteremia. The median time to all-cause mortality was 54 (IQR, 17–153) days. In survival analysis, an abnormal body mass index (BMI, normal ≥ 18.5 and <24) at the onset of bacteremia had a crude HR of 6.20 (95% CI: 1.64–23.52, $p=0.007$). The diagnosis of SLE, age, gender, DM, CKD, levels of C-reactive protein and white blood cell count did not significantly increase HR.

Table 1

Diagnosis	SLE n=2057	SjS n=1532	RA n=2364
Age (y) [median (IQR)]	40.0 (30.2–52.5)	56.3 (47.4–64.3)	56.7 (47.7–66.4)
Male gender (%)	11.3	9.1	18.3
Observation time (y) [median (IQR)]	3.22 (0.81–7.34)	1.75 (0.46–4.2)	2.77 (0.66–6.96)
NTS bacteremia (n)	26	5	2
IR per 1000 PY (95% CI)	3.10 (2.05–4.44)	1.18 (0.42–2.54)	0.22 (0.04–0.67)
Adjusted IRR (95% CI)	19.02 (4.35–83.24)**	5.72 (1.08–30.17)*	Reference
E. coli bacteremia (n)	61	15	34
IR per 1000 PY (95% CI)	7.26 (5.59–9.24)	3.54 (2.03–5.64)	3.71 (2.60–5.11)
Adjusted IRR (95% CI)	2.63 (1.67–4.16)**	0.83 (0.45–1.54)	Reference
PsA bacteremia (n)	14	4	5
IR per 1000 PY (95% CI)	1.66 (0.93–2.68)	0.94 (0.29–2.19)	0.54 (0.19–1.17)
Adjusted IRR (95% CI)	3.03 (0.98–9.35)	1.80 (0.47–6.94)	Reference

y, years; PY, person-years; *, $p<0.05$; **, $p<0.001$

Conclusion: Among the GNB bacteremia, patients with SLE were especially susceptible to NTS bacteremia when compared to RA and SjS. DM

was a risk factor. The mortality rate after NTS bacteremia was high, and an abnormal BMI may predict it.

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SAT0198 **PERIPHERAL NERVOUS SYSTEM INVOLVEMENT IN PRIMARY SJÖGREN'S SYNDROME – UNCOMMON OR UNDERESTIMATED PROBLEM?**

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Background: Systemic manifestations are common in primary Sjögren's syndrome (pSS) and play a major role in the prognosis. Neurological complications may affect both the peripheral (PNS) and central nervous system (CNS). The incidence varies from several to several dozen percent. The course of the disease and the severity of the symptoms may be mild and self-limiting or progressive, leading to permanent neurological deficits. It is worth remembering that the symptoms of nervous system involvement may precede the onset of symptoms of dryness, and diagnosis of pSS may be delayed after a certain duration of neurological symptoms.

Objectives: The aim of the study was to determine the prevalence of PNS involvement among symptomatic and asymptomatic patients with the diagnosis of pSS in our University Clinical Centre.

Methods: We studied a consecutive group of fifty unselected patients aged from 33 to 74 years (mean 55.8 years). All patients fulfilled the criteria for the diagnosis of pSS. Additional connective tissue diseases and diabetes were the exclusion criteria. Disease activity was evaluated according to the EULAR Sjögren's syndrome disease activity index (ESSDAI). All patients underwent clinical neurological examination and nerve conduction studies (NCS) of nine peripheral nerves. For the classification of polyneuropathies the ESTEEM (European Standardized Telematic tool to Evaluate Electrodiagnostic Methods) guidelines were used. Clinical examination and nerve conduction study were performed and evaluated by one certified neurologist.

Results: Of our 50 patients, 48 were female with a mean ($\pm$ SD) age 53.6 ± 10.5 years and mean disease duration 7.9 ± 5.3 years. Two were male with mean age 43.7 ± 25.8 years and mean disease duration 5.7 ± 0.6 years. The mean age at diagnosis was 50.4 ± 14 years. 23 (46%) patients fulfilled the criteria for the diagnosis of neuropathy. The most common PNS manifestation was sensorimotor neuropathy 11/23 (47%), mononeuropathy was present in 6/23 (26%) patients, pure axonal sensory neuropathy was present in 1/23 (4.3%) patient, axonal motor neuropathy in 1/23 (4.3%), SFN in 1/23 (4.3%) and cranial nerve involvement was present in 4/23 (17.4%) (one of the patients had both cranial and sensorimotor neuropathy). Neurological symptoms preceded the diagnosis of pSS in 8 (35%) of 23 PNS+ patients. The frequency of following symptoms and extraglandular manifestations was significantly higher in PNS+ compared to PNS- patients: salivary gland enlargement (74% vs 44% $p<0.05$), respiratory tract involvement (65% vs 37% $p<0.05$) and lymphadenopathy (61% vs 19% $p<0.05$). The mean ESSDAI in patients with and without neurological involvement was 7 ± 6.82 and 4.7 ± 4.58 respectively ($p=0.245$). In this subgroup the use of cyclophosphamide, due to extraglandular manifestations, was increased ($p<0.05$).

Conclusion: We found that PNS involvement is a common extraglandular manifestation of pSS (46% in our group). Sensorimotor neuropathies were most frequent. Involvement of the PNS seems frequent but remains underestimated. NCS is a non-invasive test which might be useful at diagnosis and follow-up. Guidelines for the diagnosis and treatment of peripheral neuropathies in patients with pSS are needed.

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