Systemic lupus erythematosus (SLE) is a clinically heterogeneous multi-system disease, that is characterised by the presence of autoantibodies directed against nuclear antigens. The most common manifestations include rash, arthritis, fatigue, but also anaemia, thrombocytopenia, nephritis and neurologic symptoms. Despite enormous improvements in prognosis since the introduction of immunosuppressive drugs, SLE continues to have a significant impact on the mortality and morbidity of those affected.

The aim of this project was to understand the prevalence, morbidity and outcome associated with lupus nephritis at our auto-immunity diseases centre. Clinical records of 128 patients treated between January 1993 and December 2016 were read, and national registry of Auto-immune diseases was consulted in order to characterise the Lupus cohort. Treatment was assessed and pre and post treatment biopsies were reviewed by WHO classification.

From all 137 patients assessed, average age was 49, with 93% of all patients being females. The most frequent clinical criteria were malar rash in 72%, arthritis in 51%, hematologic disturbances in 43%, from which the most frequent was lymphopenia. Among all patients, 21.2% (n=29) had clinical and histologic diagnostic criteria for lupus nephritis, with mean age at diagnosis of 34 years old (from 17 to 71). From all biopsies performed, 48% were classified as grade IV OMS. All patients were treated with glucocorticoids, and 74% performed induction therapy with Protocol Euro-Lupus, followed by mycophenolate mofetil. ACE inhibitors were used in 95.2% of all patients. Only two patients worsened and interchanged nephritis class, with one patient achieving kidney failure.

This was an important review for our centre, since our patients presented an elevated proportion of Lupus Nephritis, at a very young age.

PS9:176 SYSTEMIC LUPUS ERYTHEMATOSUS (SLE) PROSPECTIVE OBSERVATIONAL COHORT STUDY (SPOCS) TO CHARACTERISE MODERATE TO SEVERE SLE DISEASE ACTIVITY, TREATMENT, AND OUTCOMES BY TYPE I INTERFERON GENE SIGNATURE

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Purpose Type I interferon (IFN) plays a major role in SLE pathogenesis. However, limited information exists about type I IFN gene signature (IFNGS) associations with disease severity and activity, health-related quality of life, and outcomes for the general population of patients with moderate to severe SLE receiving standard-of-care treatment.

Methods Initiated in June 2017, SPOCS is an international, multicenter cohort of 1500 patients with moderate to severe SLE evaluated biannually during a 3 year follow-up period. Participating countries include Canada, United States, France, Germany, Italy, Spain, United Kingdom, and Australia. SPOCS will systematically describe the comprehensive patient journey, including clinical features, disease progression and treatment, outcomes, health status, and health care resource utilisation, for a general population of patients with moderate to severe SLE (table 1).

Association of type I IFNGS expression with these elements will be assessed. The study includes 2 year enrollment and

Abstract PS9:176 Table 1 Planned data collection

Planned Data Collection

Study site information Type of site Size of site	Treatment SLE treatment prescribed Concomitant treatment prescribed
Physician information Physician specialty Sex Number of years practicing Number of years treating patients with SLE	Other medical conditions Comorbidities Newly reported medical events
Demographics Age Sex Race and ethnicity (if allowed by local regulations) Education level Marital status Employment status	PRO questionnaires SF-36-v2 FACIT-F EQ-5D-5L LupusQoL PaGA WPAI:Lupus Medical resource use questionnaire PHQ-8
Trial participation involving investigational product or placebo (yes/no)	Pregnancy Reproductive history Pregnancy status
Physiologic variables Height/weight Blood pressure Heart rate Oral temperature Hormonal status (women)	Type I IFN gene signature
Vital status Date of death Cause of death	Biomarkers (blood sample)
SLE disease components Date of first diagnosis Date of last positive ANA/dsDNA serology ACR and/or SLICC criteria Number and severity of SLE flares SLEDAI-2K PhGA-VAS SLICC/ACR disease index score Laboratory tests Complications (including SLE-associated)	HCRU Hospitalizations Outpatient HCP visits (including surgery) Emergency department visits (nonadmitted)

ACR, American College of Rheumatology; ANA, antinuclear antibodies; dsDNA, double-stranded deoxyribonucleic acid; EQ-5D-5L, EuroQOL 5 Dimensions 5 Levels; FACIT-F, Functional Assessment of Chronic Illness Therapy-Fatigue; HCP, health care provider; HCRU, health care resource utilization; IFN, interferon; LupusQoL, Lupus Quality of Life; PaGA, Patient's Global Assessment of Disease Activity; PHQ-8, Personal Health Questionnaire Depression Scale; PRO, patient-reported outcome; SF-36-v2, Short Form Health Survey, version 2; SLE, systemic lupus erythematosus; SLEDAI-2K, Systemic Lupus Erythematosus Disease Activity Index 2K; SLICC, Systemic Lupus International Collaborative Clinics; VAS, visual analog scale; WPAI:Lupus, Work Productivity and Activity Impairment Questionnaire:Lupus.

3 year follow-up periods for each patient. Patients (≥ 18 years old) with a physician diagnosis that meets ACR or SLICC SLE criteria will be included. Additional study entry requirements include moderate to severe SLE as defined by a modified SLEDAI-2K score ≥ 4 or SLEDAI-2K score ≥ 6 , ≥ 6 month treatment duration for active SLE with systemic SLE treatment beyond NSAIDs and analgesics, and current or historic serology of ANA or dsDNA. Exclusion criteria include enrollment in interventional trials involving investigational agents or active, severe, biopsy-confirmed class III or class IV $\pm$ class V LN and/or urine protein:creatinine ratio > 1 mg/mg. Patients will be followed as per local routine clinical practice.

Results First patient recruited was achieved in June 2017, and the last patient out is anticipated for Q2 2022. Data collection, which will include use of electronic case report forms and patient-reported outcomes, will take place at biannual study visits. Distribution of type I IFNGS (test-high vs test-low) will be determined, and any association with patient outcomes will be evaluated.

Conclusion SPOCS will provide important information about possible associations of type I IFNGS with disease characteristics and outcomes for patients with moderate to severe SLE.

PS9:177 LUPUS NEPHRITIS – CLINICOPATHOLOGICAL CORRELATION AND RENAL OUTCOME

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Renal manifestation is one of the most severe complications of SLE and the clinical presentation of lupus nephritis (LN) is variable, ranging from mild asymptomatic proteinuria, renal failure to rapidly progressive glomerulonephritis. Performing renal biopsies is needed to determine the prognosis and to guide treatment in LN.

Purpose and methods To correlate the clinical, biochemical and histopathological findings in patients with biopsy proven LN and to study the renal outcome. Retrospective analysis of 100 cases of kidney biopsy proven Lupus Nephritis was done, analysed by WHO and ISN RPS Classification of Lupus Nephritis 2003.

Results 82 were female and 18 males. Extra renal manifestations in 72 cases while rest 28 had purely renal involvement. Biopsy finding showed the following class : 8 cases of class II, 29 cases of class III, 32 cases of class IV and 3 cases of class V. The remaining 28 showed combined class with predominant being class III+IV.

Significant microscopic hematuria, impaired GFR, proteinuria, anaemia, hypoalbuminemia, hypertension, and positive anti- dsDNA, low C3 were associated with the worst class, that is, class IV. Most of the patients with class IV or V had nephrotic range proteinuria and low serum albumin levels. These parameters were also correlated with high renal pathological activity and chronicity index.

24 had renal failure at the time of biopsy (mean S.creatinine 2.12 mg/dl) followed up period of 3 years. With treatment, for 12 patients renal functions improved with creatinine improving to mean value of 1.26 mg/dl. 4 patients went into severe renal failure requiring hemodialysis while 6 patients were lost to follow up.

76 patients had normal renal functions at the time of presentation with an average S.creatinine of 0.86 mg/dl. 3 patients went into ESRD. 15 patients lost follow up.

Conclusions Clinical, laboratory findings and renal biopsy are clinically valuable in identifying different renal classifications of lupus pathology, and to plan therapeutic strategies accordingly. LN class IV G was the most common and had a reduced renal survival with significant high activity and chronicity scores.

PS9:178 REMOVAL OF AN ADRENAL HORMONE ADENOMA IN A PATIENT WITH SYSTEMIC LUPUS ERYTHEMATOSUS AND AUTOMATIC FRACTURES

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Systemic lupus erythematosus is a standard autoimmune disease, characterised by the production of antibodies against cellular core. The use of corticosteroids is effective in various manifestations of lupus. We will present a 45-year-old woman suffering from the disease for 5 years and received hydroxychloroquine, prednisolone and azathioprine. The patient received bisphosphonates and after three years had an automatic right-sided fracture of pubic bone. The patient was put up in denosumab and after one year she showed a 5th metatarsal fracture left. Computed Tomography of chest and upper abdomen were performed, where it was found an adrenal adenoma. Cortisol levels in the blood were elevated. After surgery it was found that adenoma produced cortisol. Since then, there wasn't another automatic fracture.

PS9:179 THE SIDE EFFECT AND THE RETENTION RATE OF HYDROXYCHLOROQUINE IN JAPAN

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Background Hydroxychloroquine (HCQ) has been widely used in the world. However, due to the problem of retinopathy caused by chloroquine, it had not been approved in Japan until 2015, although it has been used for limited number of patients in some institutions. The primary purpose of our study is to collect the data on adverse effect of HCQ among Japanese patients with systemic lupus erythematosus. We also assessed the efficacy of our methods to improve the tolerability in cases with gastrointestinal and dermatological side effects of HCQ.

Method We retrospectively collected the data of all the 174 lupus patients treated with HCQ from the electric medical record of St Luke's International Hospital, Tokyo, Japan. We extracted the following parameters during the period between April 2008 and June 2017; patients' baseline characteristics, side effects and duration of using HCQ, and reasons of discontinuation. As for dermatological side effects, some of the patients underwent oral desensitisation; HCQ was once stopped, and after confirming negative DLST and obtaining consent from patients, it was resumed with the 36 days-desensitisation protocol. For patients with gastrointestinal side effects, some of the patients were prescribed Hangehashinto, a traditional herbal medicine for gastrointestinal discomfort, in addition to HCQ.

Result During the period, the mean age of the patients was 41.8. 161 patients (92.5%) were female and 152 patients (87.4%) were treated with steroids. The average duration of using HCQ was 582.4 days. 21 patients (12.0%) had side effects and 14 patients (8.0%) discontinued HCQ. 10 patients had skin side effects, 7 were performed DLST, 7 were negative, and 2 resumed HCQ. 5 patients had gastrointestinal side effects, 2 were prescribed Hangehashinto. 2 patients were possible retinopathy and stopped HCQ.

Conclusion This is one of the largest studies which reported well tolerability of HCQ in Japanese real world practice. Despite the concern of retinopathy, there were only 2 cases who stopped HCQ due to possible retinopathy in this study. In addition, our methods may help continuing HCQ in patients with skin or gastrointestinal side effects. Further larger study is needed to confirm these results.

PS9:180 MANAGEMENT OF NON-RETINAL TOXICITY OF HYDROXYCHLOROQUINE

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Although hydroxychloroquine is recommended for all the patients with systemic lupus erythematosus without contraindications, one third of lupus patients are not taking this important medication. Overcoming some of non-organ threatening side effects can lead to better drug survival rate and then improvement of overall outcome.

Since the approval of hydroxychloroquine in 2015 in Japan, we prescribed it to more than 200 patients of lupus in our institution. It is usually well-tolerated, but it is also true