LSO-073 IMPROVEMENTS AND CHALLENGES OF SYSTEMIC LUPUS ERYTHEMATOSUS ASSOCIATED PULMONARY ARTERIAL HYPERTENSION: A 10-YEAR MULTICENTER COHORT STUDY

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Background Previous studies have described improved survival in systemic sclerosis (SSc) associated pulmonary arterial hypertension (PAH), yet it is unclear whether survival of systemic lupus erythematosus (SLE) associated PAH has also improved.
Methods A multi-center cohort of SLE-PAH patients diagnosed by right heart catheterization (RHC) was established and divided into cohort A (2011.6-2016.5) and cohort B (2016.6-2021.5) according to the date of their baseline RHC. Another single-center cohort of idiopathic pulmonary arterial hypertension (IPAH) was consecutively recruited as control to describe the baseline characteristic and survival of SLE-PAH patients simultaneously. Disease characteristics and all-cause mortality were compared between cohort A and B. Multivariable cox regression was used to analyze association between treatment goal achievement and survival.
Results A total of 610 SLE-PAH and 104 IPAH patients were enrolled. Overall, SLE-PAH patients were younger, had lower NT-proBNP level, better function status, better hemodynamic, and higher overall survival than IPAH (81.2% vs 56.0%, p<0.001). Compared with cohort A, patients in cohort B showed lower mPAP and PVR, higher CI, and were more likely to receive extensive immunosuppressants and PAH-targeted medication. 5-year survival rate was also higher in cohort B than cohort A (88.1% vs 72.9%, p=0.01). In multivariable Cox regression, treatment goal achievement of PAH (HR 0.31, 95%CI 0.12–0.81, p=0.017) and reaching lupus low disease activity state (LLDAS) (HR 0.23, 95%CI 0.08–0.67, p=0.007) were independently associated with a lower mortality.
Conclusions This largest muti-center prospective SLE-PAH cohort showed that survival has improved significantly for SLE-PAH in the last 5 years, and for the first time, demonstrated achieving LLDAS for SLE is associated with reduced mortality for SLE-PAH patients.

LSO-074 ALL-CAUSE MORTALITY AND THE INCIDENCE OF CANCER IN PATIENTS WITH MODERATE-TO-SEVERE SLE: RESULTS FROM THE BRITISH ISLES LUPUS ASSESSMENT GROUP BIOLOGICS REGISTRY (BILAG-BR)

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Background We aimed to characterise the all-cause mortality rate (MR) and the incidence of cancer in SLE patients receiving biologic and standard of care (SoC) therapies.
Methods Patients recruited to the BILAG-BR 2010–2021 were included. Demographic and clinical data were recorded at recruitment. Mortality and malignancy data were collected from study centres, the UK Office of National Statistics and the National Cancer Register. Cox regression models were used to estimate risk in biologic-treated patients compared to SoC. Mortality models were adjusted for age, gender, co-morbidity, SLICC damage index (SDI) and hydroxychloroquine (HCQ) use.
Results During follow-up, (1463 patients with 5,962 person years [pys]), 32 incident cancers occurred in 31 individuals, a median (IQR) of 1.31 (0.63–3.36) years after registration. Compared to the UK general population, the SIR (95% CI) was 1.21 (0.85–1.72). Using SoC as the comparator, the age and gender adjusted HR was 1.49 (0.57–3.92) for rituximab and 2.47 (0.57–10.58) for belimumab. Across the whole cohort, associated risk factors (table 1), included age at recruitment (HR 1.05 [1.02- 1.08]) and male sex (HR 2.68 [1.13–6.41]).

Following registration, 54 deaths occurred after a median of 1.8 (0.8–3.3) years. The standardised MR was 4.74 (3.63–6.19). The most common causes of death were infection (22, 40.7%), SLE (11, 20.3%) and cancer (6, 11.1%). MR was 3.3 (1.3–8.9) per 1000 py in the SoC, 11.3 (8.5–15.0) in the rituximab and 2.5 (0.4–17.9) in the belimumab group. Risk factors included age at recruitment (HR 1.07 [1.05–1.09]) and SDI (HR 1.34 [1.08–1.67]. HCQ use was protective (HR 0.30 [0.14–0.65]). In multivariate analysis, compared to SoC, risk was similar in rituximab (HR 2.36 [0.69–8.10]) and belimumab groups (HR 1.41 [0.14–14.14]).
Conclusions Although overall numbers are low, mortality rate and incidence of cancer appears to be broadly similar in SoC, rituximab and belimumab treated patients.

Abstract LSO-074 Table 1 Risk factors associated with risk of cancer and death in a cohort of patientswith moderate-severe SLE

	Cancer		Death	
	Hazard ratio (HR) (95% CI)	Adjusted HR* (95% CI)	HR (95% CI)	Adjusted HR** (95% CI)
Age at recruitment (per year)	**1.07 (1.04-1.09)**	**1.05 (1.02-1.08)**	**1.07 (1.05-1.09)**	**1.04 (1.02 - 1.07)**
Male sex	**3.51 (1.57-7.86)**	**2.68 (1.13-6.41)**	**2.06 (1.01-4.22)**	1.48 (0.54 - 4.05)
Disease duration	1.02 (0.99-1.05)	-	1.01 (0.98-1.03)	-
SLICC damage index score at baseline (per unit increase)	1.20 (0.96-1.49)	-	**1.58 (1.39-1.81)**	**1.37 (1.11-1.68)**
SLEDAI at baseline (per unit increase)	0.99 (0.93-1.05)	-	1.01 (0.97-1.06)	-
Smoking ever	1.46 (0.69-3.06)	-	1.49 (0.79-2.81)	-
Previous cancer	**2.81 (1.15-6.85)**	2.05 (0.81-5.22)	**3.29 (1.58-6.82)**	2.03 (0.83-5.02)
Myocardial infarction	2.57 (0.61-10.80)	-	**5.33 (2.26-12.55)**	0.93 (0.22-4.00)
Diabetes mellitus	0.59 (0.08-4.34)	-	**3.85 (1.79-8.25)**	1.63 (0.53-4.98)
Hypertension	**2.21 (1.05-4.67)**	1.45 (0.67-3.13)	**1.81 (1.00-3.26)**	0.73 (0.32-1.67)
Ethnicity	**2.68 (1.20-5.99)**	1.77 (0.68-4.58)	**1.85 (1.05-3.26)**	0.96 (0.41-2.23)
Prednisolone dose at baseline (per mg)	0.99 (0.94-1.03)	-	0.98 (0.94-1.02)	-
Previous:				
Hydroxychloroquine	0.84 (0.32-2.18)	-	**0.33 (0.18-0.60)**	**0.31 (0.15-0.69)**
Mycophenolate mofetil	0.82 (0.39-1.69)	-	1.16 (0.66-2.07)	-
Cyclophosphamide	1.30 (060-2.83)	-	1.1 (0.65-2.14)	-

*Adjusted for age, sex, previous cancer, hypertension and ethnicity

**Age, sex, previous cancer, myocardial infarction, diabetes mellitus, hypertension, white ethnicity and HCQ use