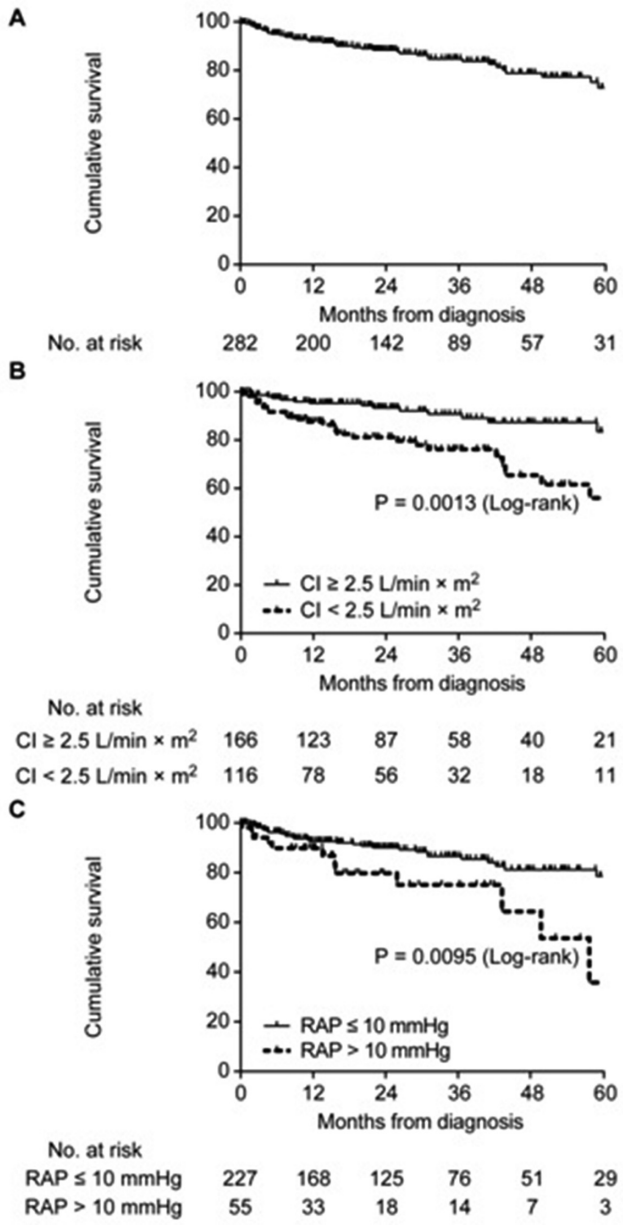
follow-up data were included in the TGA study. The median follow-up was 24.0 months. The 1-, 3- and 5 year survival rates were 92.1%, 84.8% and 72.9%, respectively. The 1-, 3- and 5 year TGA rates were 31.5%, 53.6% and 62.7%, respectively. Serositis (HR=1.94, 95% CI: 1.26–3.00, p=0.003), 6MWD >380 m (HR=1.95, 95% CI: 1.14–3.31, p=0.014) and CI ≥2.5 L/min×m^2 (HR=1.92, 95% CI: 1.16–3.19, p=0.012) 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.

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. PAH patients with high SLE activity may benefit from immunosuppressive therapy.



Abstract 242 Figure 1

243 THE PIVOTAL ROLE OF INTENSIVE IMMUNOSUPPRESSIVE THERAPY IN THE MANAGEMENT OF SYSTEMIC LUPUS ERYTHEMATOSUS ASSOCIATED WITH PULMONARY ARTERIAL HYPERTENSION

[1]J Zhao*, [1]Q Wang, [1]J Qian, [2]Z Tian, [2]Y Liu, [1]M Li, [1]X Zeng. *[1]Peking Union Medical College Hospital, Rheumatology, beijing, China; [2]Peking Union Medical College Hospital, Cardiology, beijing, China*

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Background and aims Immune and inflammatory mechanisms could play a significant role in pulmonary arterial hypertension (PAH) genesis and progression, especially in patients with systemic lupus erythematosus (SLE). Immunosuppressive therapy should be better evaluated in this setting. We reviewed the clinical outcomes of intensive immunosuppressive therapy with or without target therapy in SLE associated P

Methods This single-centre cohort study enrolled 126 consecutive patients with SLE-PAH who visited our referral centre in China between May 2006 and December 2015. Baseline demographics, clinical features, laboratory results, haemodynamic assessments and management were analysed. Kaplan-Meier curves and Cox proportional hazards regression analysis were used toevaluate the role of intensive immunosuppressive therapy.

Results ALL patients received intensive immunosuppressive therapy including combination of high-dose glucocorticosteroids and first-line immunosuppressants, such as cyclophosphamide, mycophenolate and calcineurin Inhibitors. Eighty-two (65.1%) patients received target therapy at baseline. Survival analysis indicated that responders had a better survival than nonresponders in both with and without target therapy group (figure 1). Patients with a shorter SLE disease duration (p=0.009) and better baseline pulmonary hemodynamics (mean pulmonary arterial pressure, pulmonary vascular resistance and Cardiac index, p<0.001) were more likely to benefit from immunosuppressive therapy (table 1).

Conclusions Intensive immunosuppressive therapy markedly improved the long-term outcomes of SLE patients with PAH, especially in the early stage of PAH.

244 LONG-TERM PROGNOSIS AND PREDICTING FACTORS OF CHINESE PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS: A MULTI-CENTRECENTER COHORT STUDY FROM CSTAR REGISTRY

[1]J Zhao*, [1]Z Wang, [2]Y Wang, [1]Q Wang, [1]M Li, [1]X Zeng. *[1]Peking Union Medical College Hospital, Rheumatology, beijing, China; [2]School of Basic Medicine Peking Union Medical College and Institute of Basic Medical Sciences Chinese Academy of Medical Sciences, Department of Epidemiology and Biostatistics, beijing, China*

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Background and aims To investigate the long-term outcomes, both mortality and damage, and predict factors of patients with systemic lupus erythematosus (SLE) in the CSTAR (Chinese SLE Treatment and Research group) registry cohort.

Methods Patients were enrolled from April 2009 to February 2010. They were followed up at clinic and were telephone interviewed at the endpoint. Demographic data, clinical manifestations, activity, damage scores, and medications were collected. Survival rates were studied by Kaplan-Meier method,

Abstract 243 Table 1 Comparison of clinical characteristics in responders and nonresponders to immunosuppressive therapy

	SLE-PAH without target therapy			SLE-PAH with target therapy		
	Responder N=29	Nonresponder N=15	*p-value*	Responder N=44	Nonresponder N=38	*p-value*
Female, n, (%)	29(100)	15(100)	*1.000*	43(100)	37(97.4)	*1.000*
Age, years	33.8±9.2	37.0±10.0	*0.293*	32.1±7.2	35.3±8.3	*0.066*
SLE Disease duration, months	3.5(0,23.7)	6.4(1.0,33.1)	*0.090*	4.8(0,18.9)	6.3(0.7,23.1)	*0.427*
RP,n(%)	19(65.5)	9(60.0)	*0.718*	24(54.5)	24(63.2)	*0.430*
Anti-u1RNP, n (%)	21(72.4)	10(66.7)	*0.676*	25(61.0)	19(50.0)	*0.326*
SLEDAI-2000	7.0±6.2	5.1±4.5	*0.296*	3.0±2.9	3.5±2.6	*0.420*
WHO functional classification						
I-II, n(%)	17(58.6)	5(33.3)	*0.013*	21(47.7)	18(47.6)	*0.292*
III-IV, n(%)	12(41.4)	10(66.7)		23(52.3)	20(52.6)	
6MWD, meter	465.3±77.4	424.0±97.9	*0.180*	417.8±99.4	398.2±92.9	*0.409*
Mean RAP, mmHg	4.2±3.0	2.8±2.8	*0.201*	3.9±4.3	4.3±4.0	*0.706*
Mean PAP, mmHg	37.9±8.2	45.7±7.9	*0.005*	45.1±10.3	53.2±11.0	*0.001*
CI, $l.min^{-1}.m^{-2}$	3.2±0.7	2.5±0.6	*0.003*	2.8±0.6	2.4±0.8	*0.018*
PVR, WU	6.6±2.4	10.5±3.0	*0.001*	9.2±3.6	12.7±4.5	*0.000*

and COX proportional hazard model was adopted to perform the analysis of predicting factors for mortality.

Results A total of 2104 patients were recruited at baseline, and 1494 patients were successfully followed up. The cumulative 1, 3 and 5 year survival rates from diagnosis were 99.0%, 98.1% and 97.1%. 78 patients died during follow-up, and the main death causes were infection (34.6%), active disease (26.9%), cardiovascular and cerebrovascular events (6.41%) and malignancy (5.13%). At entry, 247 patients presented with irreversible organ damage, 398 patients at the endpoint. The major accumulated organ damages were renal (25.9%), musculoskeletal (20.2%), neuropsychiatric (12.4%), and pulmonary (10.8%) damage. Cox regression showed that male, late onset (≥50y), onset to diagnosis time ≥1 year, previous organ damage, renal involvement, pulmonary arterial hypertension, neuropsychiatric involvement, serositis and the number of involved organ systems ≥3predict for higher mortality.

Conclusions Long-term survival rates of SLE patients have been improved in China. Early diagnosis, preventing from the emerging systemic organ involvements and organ damage could be the treating target for the management of SLE patients.

Clinical trials

245 HIGH DOSE OF VITAMIN D THERAPY AND URINARY ANGIOSTATIN AMONG EGYPTIANS JUVENILE LUPUS

S Algergawy [1]*Benha university faculty of medicine, rheumatolgy and rehabiliatation department, Nasr City, Egypt*

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Background and aims Vitamin D has numerous effects on cells within the immune system. Association between vitamin D deficiency and high disease activity in systemic lupus was confirmed. The aim of this research was to study the effect of vitamin D