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

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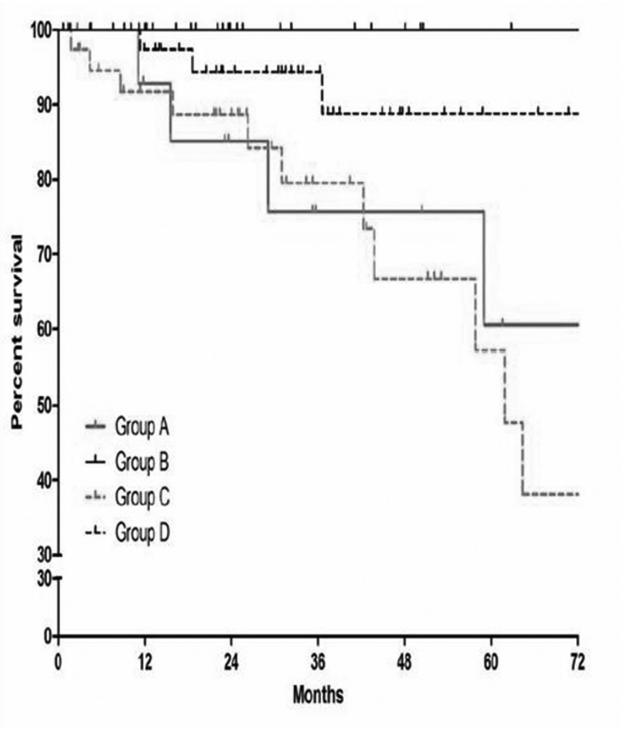
Background and aims Vitamin D has numerous effects on cells within the immune system. Association between vitamin D deficiency and high disease activity in systemic lupus was confirmed. The aim of this research was to study the effect of vitamin D



Notes: Group A=SLE-PAH without target therapy and responder; Group B=SLE-PAH without target therapy and nonresponder; Group C=SLE-PAH with target therapy and responder; Group D=SLE-PAH with target therapy and nonresponder;

Abstract 243 Figure 1 Survival of patients with SLE-PAH treated with intensiveimmunosuppressive therapy with or without PAH-target therapy

therapy on urinary angiostatin as a marker of activity in juvenile lupus.

Methods Fifty female patients with juvenile systemic lupus erythematosus (SLE) were enrolled in this study for twelve weeks trial of high dose of oral vitamin D weekly. We used radioimmunoassay to measure Serum levels of vitamin D. Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) and the European Consensus Lupus Activity Measurement (ECLAM) were measured to assess lupus activity. Urinary angiostatin was evaluated as marker of activity. All parameters were measured on the day one of study and after three months. Thirty juvenile SLE patients as control.

Results At beginning of study all patients and control were almost similar regarding age, clinical, laboratory, urinary angiostatin and vitamin D levels. After three months the mean Vitamin D level was increase in patients group received Vitamin D than in control group ($p<0.001$). There was significant decrease in urinary angiostatin ($p<0.05$) was observed in the patients received vitamin D compared to patient without vitamin D supplementation.

Conclusions Our findings show that high dose of vitamin D supplementation up to 12 weeks is safe and diminish lupus activity. We need longer duration and more studies to confirm our results.

246 HIGH TITER ANA WITH ANTI-DFS70 ALONE IS NOT TO BE CONSIDERED A VALID CRITERION FOR LUPUS

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Background and aims Positive ANA is one of Criteria for Classification of SLE for ACR and SLICC. As a follow-up to the International Consensus on ANA Patterns (ICAP) initiative (ANApatterns.org), the relevance of each ANA pattern is being re-evaluated.

Methods ANA test at 1/80 screening dilution was performed in 269 sequentially selected patients with SLE diagnosis, 918 healthy individuals, and 558 patients with non-SARD conditions. ANA interpretation was the consensus of 3 independent readers using 2 HEp-2 cell slide brands at 400x mag. Conversely, sequentially selected individuals presenting >1/640 titer Nuclear Dense Fine Speckled (DFS) ANA pattern (AC-2) in a large clinical laboratory within a 2 year period had the diagnosis assessed by interview with the respective physician.

Results Among 269 consecutive SLE patients, 96.3% had a positive ANA with the following principal nuclear patterns: homogeneous (29.3%), coarse speckled (14.7%), fine speckled (40.1%). Only one patient (0.3%) had the DFS pattern and the reactivity to DFS70 confirmed by ELISA. Conversely, among 118 ANA+ healthy individuals and 102 ANA+ patients with miscellaneous non-SARD conditions, 33% and 17% presented the DFS pattern, respectively. In addition, the 327 consecutive high-titer DFS individuals presented mostly non-SARD conditions or non-specific clinical presentation. Only 7 had possibly SARD-related presentations: 1 anti-phospholipid syndrome, 1 "possible" SLE (polyarthritis, arthritis, chronic urticaria), 1 WG, 1 DLE, 1 PBC, and 1 RA.

Conclusions Well-defined anti-DFS ANA, confirmed by antigen-specific reflex testing, should not be considered a criterion for SLE - either in the ACR or SLICC classification criteria.

247 THE EFFICACY OF ANTI-CD20 ANTIBODY RITUXIMAB FOR REFRACTORY PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background and aims B cells play a crucial role in pathogenesis of Systemic Lupus Erythematosus(SLE). We examined the efficacy of B cell depletion therapy rituximab for refractory patients with SLE.

Methods 63 eligible study subjects since 2002 until 2015 were men and women, who met the American College of Rheumatology criteria in 1987 or SLICC2012 for the classification of SLE. The protocols were approved by the Institutional Review Board of our university. Treatment protocol: 2 or 4 weekly doses of 500 mg/body, 2 biweekly doses of 1000 mg/body or 4 weekly doses of 1000 mg/body.

Results Baseline characteristics; gender M:F=6:57, age 33.9 years, disease duration 87.2 months, organ failure NPSLE:35, lupus nephritis:46, treated with IVCY 34/63. The 60/63