lupus anticoagulant positivity, nor with single-, double- or triple- APLA positivity.

Conclusion Our data show for the first time that plasma sTREM-1 level is significantly elevated in patients with thrombotic PAPS. We suggest that soluble TREM-1 might be used as a biomarker for thrombosis in patients with primary APS and our results support the possible role for the innate immune system in the pathogenesis of thrombosis in PAPS.

S3D:6 MICROANGIOPATHIC MANIFESTATIONS OF THE PRIMARY ANTIPHOSPHOLIPID SYNDROME

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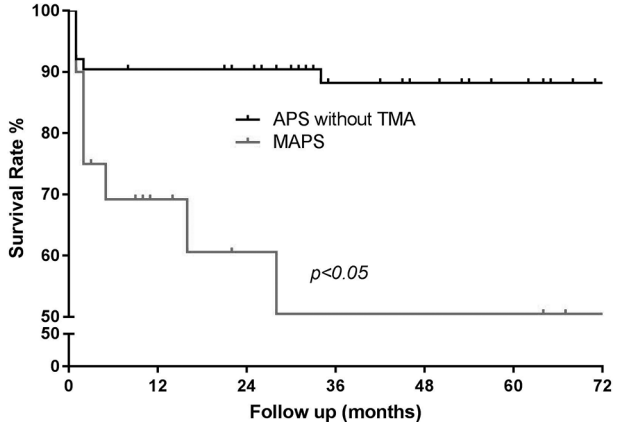
Objective Thrombotic microangiopathy (TMA) is a relatively rare but severe vascular complication of antiphospholipid syndrome (APS) and may involve any organ. It's crucial for prompt diagnosis and early intervention, while the clinical features and outcomes of patients with TMA in APS is quite scant. The aim of this study was to investigate characteristics of primary APS with TMA patients.

Patients and methods One handred and twelve priamry APS patients who attended in Peking Union Medical College Hospital from January 2004 to December 2016 were enrolled. Patients were subdivided into two groups: PAPS with TMA and PAPS without TMA. Demographic data, clinical characteristics, laboratory features and treatment were retrospectively collected. Bivariate statistical analysis and logistical regression test were performed to compare the discrepancy between the two groups. Survival rates were studied by Kaplan-Meier method, and COX proportional hazard model was adopted to perform the analysis of predicting factors for mortality.

Results Twenty-one patients with TMA were indentified, presenting in 18.8% of all PAPS patients. Both groups were similar with respect to age of onset, gender, clinical course and pregnancy morbidity. However, the frequency of anti-b2-glycoprotein I antibodies (anti-b2GPI) (66.7% vs 33.0%, p=0.004) was significantly higher in the TMA group than the non-TMA group. In addition, thrombocytopenia (85.7% Vs 54.9% p=0.007) and elevated high-sensitivity C-reactive protein (hsCRP) (71.4% Vs 54.9% p=0.007) were observed more frequently among TMA group. In multivariate analysis, only thrombocytopenia (OR=4.055, 95% CI: 1.006 to 16.345, p=0.049), hsCRP elevation (OR=6.789, 95% CI: 2.018 to 22.840, p=0.002) and positivity for anti-b2GPI (OR=4.723, 95% CI: 1.409 to 15.830, p=0.012) were independent risk factors for the occurrence of TMA in PAPS patients. The overall 1, 3, and 5 year survival rate of PAPS with TMA was 69.2%, 60.6% and 50.5% respectively; while 90.4%, 88.2%, and 88.2% in APS patients without TMA (p<0.05).



Abstract S3D:6 Figure 1

Conclusion Microangiopathic antiphospholipid syndromes should be an independent subset with mainly small vessels involved and worse prognosis, compared with classic APS.

S3D:7 CEREBRAL HYPOPERFUSION DETECTED BY PERFUSION-WEIGHED MRI MAY ASSIST THE DIAGNOSIS OF PRIMARY DIFFUSE NEUROPSYCHIATRIC LUPUS ERYTHEMATOSUS

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Objective Abnormalities in regional or global cerebral perfusion have been reported in SLE but their value in distinguishing lupus from non-lupus related diffuse neuropsychiatric events has not been determined. We examined whether the addition of dynamic susceptibility-contrast-enhanced T2*-weighted perfusion MRI (DSC-MRI), a non-invasive assessment of brain haemodynamic status, to the standard MRI examination suggested by the EULAR recommendations, may be of added value in the clinical diagnosis and attribution of neuropsychiatric SLE (NPSLE).

Patients and methods Seventy-six SLE patients (53 NPSLE, 23 non-NPSLE) and 31 healthy controls underwent conventional MRI (cMRI) and DSC-MRI. Attribution of NPSLE to lupus (primary NPSLE: n=37) or not (n=16) was based on multidisciplinary assessment including cMRI results and response to treatment. Cerebral blood volume (CBV) and cerebral blood flow (CBF) values were estimated in 18 normal-appearing white matter (NAWM) and deep grey matter (NADGM) areas. Perfusion differences among subgroups and their diagnostic utility were assessed using Analysis of Variance, Receiver Operating Characteristics, and Binary Logistic Regression analysis.

Results The most common manifestations were mood disorder, cognitive disorder and headache. The most common manifestations were mood disorder, cognitive disorder and headache. Primary NPSLE patients had lower cerebral blood flow and volume in several NAWM areas compared to controls (p<0.0001), and lower cerebral blood flow in the semioval centre bilaterally compared to non-NPSLE and non-primary NPSLE patients (p<0.001). A cut-off for cerebral blood flow of 0.77 in the left semioval centre discriminated primary NPSLE from non-NPSLE/non-primary NPSLE with 80% sensitivity and 67%–69% specificity. Blood flow values in the left semioval centre showed substantially higher sensitivity than cMRI (81% versus 19%–24%) for diagnosing primary NPSLE and the combination of the two modalities yielded 94%–100% specificity in discriminating primary from non-primary NPSLE.

Conclusion Primary NPSLE is characterised by significant hypoperfusion in white matter and deep grey matter areas that appear normal on conventional MRI sequences. Addition

of cerebral blood flow in the semioval centre to conventional MRI techniques described in the EULAR NPSLE recommendations improves the diagnosis of primary NPSLE.

S4a – Longterm outcome

S4A:4 BETA2-MICROGLOBULIN (B2MG) PLASMA LEVELS ASSOCIATE WITH MARKERS OF ATHEROSCLEROSIS IN PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

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Purpose SLE is associated with premature and accelerated atherosclerosis. Atherosclerotic processes are associated with impaired renal function and may be driven by inflammatory pathways, including interferon type 1 responses; both conditions with relevance to SLE. Plasma levels of B2MG are correlated to interferon-alpha activity in SLE as well as dependent of renal function. In this study plasma B2MG was correlated to markers of atherosclerosis in patients with SLE.

Methods Patients with SLE (n=147) were included in the cross-sectional study. Traditional cardiovascular risk factors were accounted for. Markers of atherosclerosis included 1) coronary artery calcium (CAC) assessed by computed tomography without contrast and identified in accordance with the Agatston scoring method and 2) carotid plaque (CP) assed by ultrasound and identified as either 1) a local thickening of the intima-media thickness of >50% compared with the surrounding vessel wall, 2) an IMT of more than 1.5 mm thick or 3) a local thickening of the arterial wall of more than 0.5 mm; bilateral examination. P-B2MG was measured by a routine immunoturbidimetric assay. The Cockgroft-Gault formula was used to calculate the estimated glomerular filtration rate (eGFR) (mL/min/1.73 m^2 body surface area) which was stratified into quartiles.

Results CAC, CP or any of them (CAC/CP) were found in 57, 29 and 62 patients, respectively; eGFR <90 was found in 74 patients. P-B2MG having a median of 216 nmol/L (range: 101–2810). Among the patients with the highest quartile of P-B2MG the frequency of CAC/CP was around 65% irrespective of eGFR. However, in a full logistic regression model taking into account traditional and disease related cardiovascular risk factors, patients with normal eGFR and a high quartile P-B2MG where found to have the highest odds ratio for having CAC/CP, see table 1.

Abstract S4A:4 Table 1

	SLE patients with eGFR <90 (n = 74)		SLE patients with eGFR ≥90 (n = 73)	
Variable	OR (95% CI)	P-value	OR (95% CI)	P-value
P-B2MG				
- quartile 1	0.96 (0.10–9.07)	0.97	1	-
- quartile 2	0.72 (0.14–3.73)	0.70	2.12 (0.41–11)	0.37
- quartile 3	3.32 (0.63–17)	0.16	1.04 (0.16–6.59)	0.99
- quartile 4	3.89 (0.80–19)	0.09	15.45 (1.97–121.45)	0.01

Conclusion We found a high prevalence of atherosclerotic markers in patients with SLE with the highest among those within the top quartile of P-B2MG in combination with normal renal function. These results suggest that atherosclerosis in SLE may be associated with interferon-alpha activity irrespective of renal function.

S4A:5 HIGH GENETIC RISK SCORE IS ASSOCIATED WITH ORGAN DAMAGE IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background Systemic lupus erythematosus (SLE) is a chronic, autoimmune disease with a complex genetic aetiology. The overall effect of hereditary risk on organ damage has so far not been studied. We therefore assessed the relationship between genetic risk and development of organ damage in SLE.

Methods Patients with SLE (Sweden, n=1001) were genotyped using a 200K Immunochip single nucleotide polymorphism (SNP) Array (Illumina). The Immunochip was HLA imputed using HLA*IMP:02. A non-HLA (58 SNPs) and a HLA (5 SNPs) genetic risk score (GRS) was assigned to each patient based on SNPs which in previous studies have shown association ($p<5\times10–8$) with SLE in European populations (Chen *et al*. 2017). For each SNP, the natural logarithm of the odds ratio (OR) for SLE susceptibility was multiplied by the number of risk alleles in each individual. The sum of all products for each patient was defined as the GRS. Clinical data, including the Systemic Lupus International Collaborating Clinics Damage Index (SLICC-DI), was retrieved from medical records. The relationship between GRS and SLICC-DI was analysed using an ordinal regression model.

Results A higher non-HLA GRS was associated with increased organ damage (OR 1.10 (1.00–1.21), $p=4.2\times10–2$), nephritis (OR 1.26 (1.13–1.41), $p=2.8\times10–5$), anti-dsDNA (OR 1.33 (1.17–1.52), $p=1.0\times10–5$) and a younger age at diagnosis (OR 1.33 (1.14–1.54), $p=1.7\times10–4$).

When analysing the relationship between individual SNPs (n=63) and SLICC-DI, we observed positive associations between SLICC-DI and rs6568431 (ATG5, OR 1.28 (1.08–1.51), $p=3.6\times10–3$) and rs11889341 (STAT4, OR 1.27