Conclusion A strong association was observed between anti-dsDNA antibodies measured using Aptiva dsDNA PMAT and renal involvement and between anti-Sm antibodies and neurological manifestations. The presence of anti-dsDNA, anti-Sm and anti-Ribo-P autoantibodies were higher in patient with renal involvement.

PO.2.36 CHARACTERIZATION OF PERIPHERAL BLOOD B-CELL SUBSET IN UNTREATED PATIENTS WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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Background B-cells play a critical role in the regulation of systemic autoimmune diseases pathogenesis, that extends beyond antibodies production.
Objectives To examine B-cells subsets in peripheral blood of patients (pts) with untreated systemic lupus erythematosus (SLE). To analyze the association between B-cell subsets and disease-specific autoantibodies.
Methods Thirty six untreated SLE pts (31F/5M) were enrolled. SLE pts had median age of 37 years (range) (32–41), disease duration of 3.5 years (1–11), SLEDAI 2K of 7 (4–8), BILAG of 14,5 (10,2–23).

The control group consisted of 29 volunteers (23F/6M, median age 38 [35; 48] years). CD19+B cells, memory B cells (CD19+CD27+), switched memory B cells (CD19+ CD27 +IgD-), non-switched memory B cells (CD19+CD27+ IgD+), naive (CD19+CD27-IgD+), double-negative (CD19+CD27-IgD-), transitional (CD19+CD38++CD10+IgD+CD27-) B cells, and plasmablasts (CD19+CD38+++CD27+IgD-CD20-) were analyzed using multicolor flow cytometry.
Results The absolute counts of memory B cells (CD19+CD27 +), switched memory B cells (CD19+CD27+IgD-), transitional B cells (CD19+CD38++CD10+IgD+CD27-), and plasmablasts (CD19+CD38+++CD27+IgD-CD20-) were higher in untreated pts with SLE compared to healthy donors, p<0,01 for all cases. The absolute counts of double-negative B cells (CD19+CD27-IgD-) were lower in SLE pts than in donors, p=0,03 (Table 1).

Abstract PO.2.36 Table 1 Levels of the blood B-cell subsets in SLE pts and in control

Parameters, n (x10^9/l)	SLE (n=36)	Control group (n=29)
CD19+B cells	0,15 (0,12-0,25)	0,1 (0,08-0,2)
memory B cells (CD19+CD27+)	0,036(0,031-0,006) *	0,003(0,001-0,007) *
switched memory B cells (CD19+ CD27+IgD-)	0,02 (0,016-0,046) *	0,01 (0,005-0,02) *
non-switched memory B cells (CD19+CD27+ IgD+)	0,012(0,007-0,027)	0,02(0,01-0,04)
naive B cells (CD19+CD27-IgD+)	0,09 (0,06-0,2)	0,1 (0,06-0,1)
double-negative B cells (CD19+CD27-IgD-)	0,011 (0,006-0,014) *	0,02 (0,01-0,02) *
transitional B cells (CD19+CD38++CD10+IgD+CD27-)	0,011 (0,004-0,026) *	0,0001 (0-0,0003) *
plasmablasts (CD19+CD38+++CD27+IgD-CD20-)	0,001 (0,001-0,002) *	0,0001 (0,00006-0,0003) *

Note: * - Differences in B-cell subset between SLE and control groups.

At significant correlation was found in SLE pts between anti-dsDNA levels and absolute counts of the following B-cell subtypes: CD19+B cells (r=0,72), memory B cells (CD19 +CD27+) (r=0,76), switched memory B cells (CD19+ CD27 +IgD-) (r=0,75) and naive B cells (CD19+CD27-IgD+) (r=0,73), p<0,01 for all cases. In addition, in SLE pts the a-Sm levels correlated with absolute counts of the switched memory B cells (CD19+ CD27+IgD-) (r=0,98; (r=0,51, p<0,01), antibodies to cardiolipin (aCL) IgG levels with absolute counts of CD19+B cells (r=0,64, p=0,02), and with naive B cells (CD19+CD27-IgD+) (r=0,59, p=0,01).
Conclusions Immunophenotyping showed an increase in the absolute counts of memory B cells (CD19+CD27+), switched memory B cells (CD19+CD27+IgD-), transitional B cells (CD19+CD38++CD10+IgD+CD27-), and plasmablasts (CD19+CD38+++CD27+IgD-CD20-) in untreated SLE compared with healthy subjects. Positive correlation between the counts of B-cells subsets and values of disease-specific autoantibodies (anti-dsDNA, a-Sm, aCL IgG) suggests that B-lymphocytes may be involved in SLE pathogenesis.

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PO.2.37 RISK FACTORS FOR RECURRENT THROMBOSIS AND CAUSE OF DEATH IN PATIENTS WITH ANTIPHOSPHOLIPID SYNDROME; A SWEDISH COHORT STUDY

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Background In patients with the antiphospholipid syndrome (APS), recurrent thrombosis (re-thrombosis) is common despite anticoagulation and the mortality rate is high. Concomitant systemic autoimmune rheumatic diseases (SARD) are frequent in patients with APS and often associated with disease associated damage. Less is known about the prevalence of non-rheumatic autoimmune diseases (NRAID) in these patients.
Purpose To estimate the incidence of re-thrombosis and death, evaluate the impact of cardiovascular (CV) risk factors and antiphospholipid antibody (aPL) profiles on re-thrombosis and identify causes of death in a novel APS cohort. To evaluate the incidence and prevalence of concomitant autoimmune diseases (AID) in this cohort.
Methods This retrospective cohort study comprises all patients identified with APS in the electronic medical records at Karolinska University Hospital, Sweden 2014–2020. Descriptive statistics was presented as median and interquartile range (IQR). Cox proportional hazards regression analyses were used to investigate the effect of risk factors.
Results 271 patients were included in the cohort. Age of APS-diagnosis was 43 years (IQR 31–55) and 66% were women. At inclusion, 130 (48%) patients presented with AID; 101 (37%) had a concomitant SARD while 54 (19%) had a NRAID. Systemic lupus erythematosus was the most frequent

in 30%, followed by autoimmune thyroid disease in 10% of patients. During follow-up, 37 re-thrombosis occurred; 23 arterial and 14 venous events, with an incidence of 3.4 per 100 person-years (95% CI: 2.4–4.7). Significant CV risk factors for re-thrombosis were current smoking; hazard ratio 2.50, p=0.03 and chronic kidney disease; 3.44, p <0.01. Twenty-seven (73%) patients with re-thrombosis were triple positive for aPL compared to 113 (48%) without any event at follow-up (p <0.01). The cumulative death incidence was 4% (n=12) with sepsis due to bacterial infection being the most common cause. The median age at death was 63 years (IQR 51–71) and occurred 8 years (IQR 2–10) after diagnosis.

In addition, 35 (13%) APS-patients developed AID during the study period, corresponding to an incidence rate of 28.4 (95% CI; 19.3–40.3) per 1.000 person-years with mean time at risk of 4 (SD±2) years.

Conclusion APS patients are at high risk to develop other AID during the 5 years follow-up period. Multiple CV risk factors are present in APS-patients suffering re-thrombosis, with smoking and chronic kidney disease being most important. APS-patients are susceptible to sepsis following bacterial infection with high mortality. These findings might be helpful when considering risk stratification and alternate treatment options in this patient group.

PO.2.38 ANTI-β2GPI-DOMAIN 1 ANTIBODIES STRATIFY HIGH RISK OF EXTRA-CRITERIA MANIFESTATIONS IN A LARGE PROSPECTIVE CHINESE COHORT OF ANTIPHOSPHOLIPID SYNDROME

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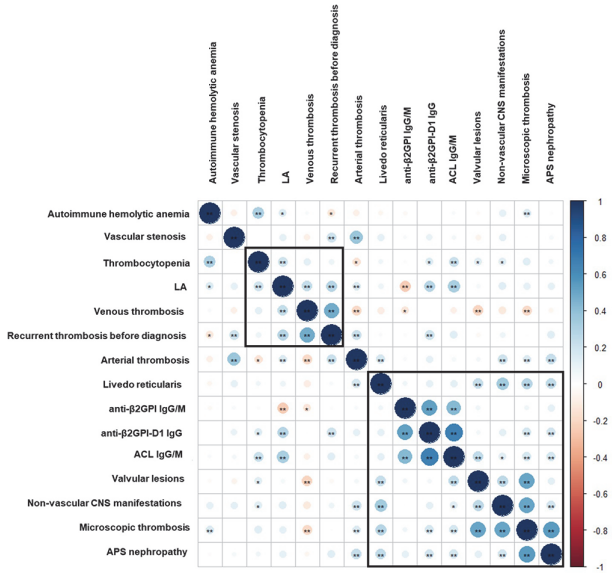
Purpose Anti-β2GPI-Domain 1 (β2GPI-D1) antibodies are potentially pathogenic in patients with antiphospholipid syndrome (APS), but their clinical associations were unclear. We aimed to evaluate the clinical characteristics of APS patients with anti-β2GPI-D1 positivity, and its utility in diagnosing APS among SLE patients.

Methods A total of 338 patients were included, of which 169 patients diagnosed with primary APS (PAPS group), 50 with APS secondary to SLE (SAPS group), 209 with SLE (SLE group). Serum anti-β2GPI-D1 IgG was measured using chemiluminescent immunoassay (Inova Company). Extra-criteria manifestations were analyzed, including thrombocytopenia, autoimmune hemolytic anemia, valvular lesions, APS nephropathy, and non-vascular neurological manifestations.

Results Similar presence of anti-β2GPI-D1 IgG was seen among PAPS (32.80%) and SAPS (32.0%) patients, and 96.4% of those with positive anti-β2GPI-D1 IgG showed triple aPLs positivity. Anti-β2GPI-D1 IgG was significantly associated with recurrent thrombosis before APS diagnosis, microscopic thrombosis (p<0.05), but not with adverse pregnancy events (Figure 1). Notably, patients with extra-criteria manifestations, especially thrombocytopenia and APS nephropathy, showed significantly higher titers in anti-β2GPI-D1 IgG (p<0.05). After a median follow-up of twenty-five months, patients with anti-β 2GPI-D1 IgG also showed a tendency of more extra-criteria events (3/55 vs 1/114, p=0.095), but not thrombotic events or adverse pregnancy events. Anti-β2GPI-D1 was positive



Abstract PO.2.38 Figure 1

among 8.13% of the SLE controls, and showed higher specificity (91.9%) in diagnosing SAPS among SLE patients as compared to classic aPLs.

Conclusions Anti-β2GPI-D1 IgG had a stronger association with extra-criteria manifestations in APS patients compared to three classic APLs, which properly indicated its pathogenic role of microangiopathy.

PO.2.39 LONG TERM FOLLOW UP OF PATIENTS WITH PRIMARY OBSTETRIC ANTIPHOSPHOLIPID SYNDROME

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Introduction Primary obstetric antiphospholipid syndrome (OAPS) is defined by specific morbidities and/or losses of pregnancy in the presence of persistent antiphospholipid antibodies (aPL). This variant of APS is usually treated during pregnancy and the post-partum period. Data on occurrence of thrombotic event during long term follow-up of OAPS patients is limited.

Methods A multi-center retrospectively cohort of female patients with primary APS (pAPS) was assembled during 2004–2019. Patients were grouped according to disease presentation as pure OAPS or thrombotic APS (tAPS) for those presenting with thrombosis. Clinical and serological data were compared between groups.

Results Of 219 pAPS female patients 67 (30.6%) were diagnosed with OAPS and 152 (69.4%) with tAPS. During >10 years of follow-up 24/67 (35.8%) OAPS and 71/152 (50%) tAPS suffered a new thrombotic event (p = 0.06), while obstetric morbidity was more likely in the OAPS group (31.3 vs. 10.5%, p < 0.001) respectively. Among patients with OAPS at presentation heart valve disease and the presence of