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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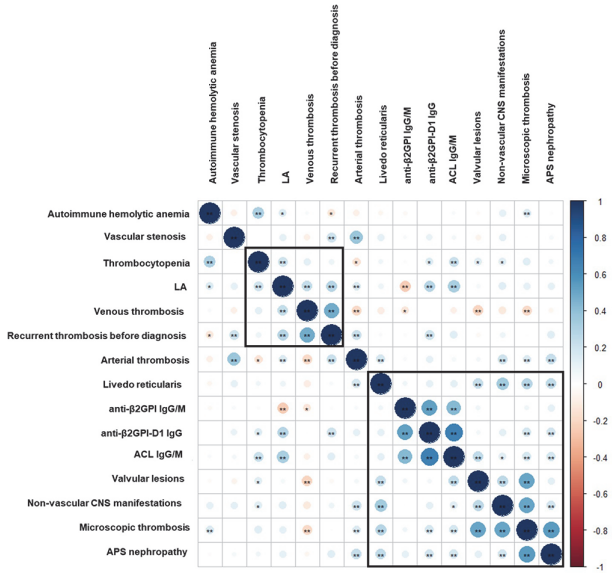
Purpose Anti-β2GPI-Domain 1 (β2GPI-D1) antibodies are potentially pathogenic in patients with antiphospholipid syndrome (APS), but their clinical associations were unclear. We aimed to evaluate the clinical characteristics of APS patients with anti-β2GPI-D1 positivity, and its utility in diagnosing APS 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 among 8.13% of the SLE controls, and showed higher specificity (91.9%) in diagnosing SAPS among SLE patients as compared to classic aPLs.



Abstract PO.2.38 Figure 1

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