P9 VALIDATION OF THE ADJUSTED GLOBAL ANTIPHOSPHOLIPID SYNDROME SCORE AND CORRELATION WITH EXTRA-CRITERIA MANIFESTATIONS

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Background/Purpose Adjusted global antiphosholipid syndrome score (aGAPSS) is the simplified version GAPSS that was recently developed to assess thrombotic risk by the consideration of antiphospholipid antibody (aPL) profile and conventional cardiovascular risk factors. The aim of this study was to evaluate the validity of the aGAPSS in predicting thrombosis and extra-criteria manifestations in our antiphospholipid syndrome (APS) cohort.

Abstract P9 Table 1 Demographic, laboratory and clinical characteristics of patients

	Total population (*n*=98) *n* (%)	PAPS (*n*=42) *n* (%)	SLE/APS (*n*=56) *n* (%)	*P*
Female	83 (84.7)	36 (85.7)	47 (83.9)	0.52
Age, years (mean±SD)	42.4 (10.9)	44.6 (11.6)	40.8 (10.1)	0.42
Disease duration, years (mean±SD)	9.8 (7.8)	10 (8.8)	9.7 (7.1)	0.16
Thrombosis	87 (88.8)	35 (83.3)	52 (92.9)	0.12
• Arterial	58 (66.7)	24 (68.6)	34 (65.4)	0.47
• Venous	45 (51.7)	19 (54.3)	26 (50)	0.43
• Recurrent	37 (42.5)	15 (42.9)	22 (42.3)	0.56
o A→V	3 (8.1)	2 (13.3)	1 (4.5)	
o V→A	15 (40.5)	6 (40)	9 (40.9)	
o A→A	9 (24.3)	2 (13.3)	7 (31.8)	
o V→V	10 (27)	5 (33.3)	5 (22.7)	
Pregnancy morbidity	40 (40.8)	20 (47.6)	20 (35.7)	0.16
• <10 weeks, ≥ 3 abortions	9 (22.5) 29 (72.5)	5 (25) 14 (70)	4 (20) 15 (75)	0.5 0.5
• ≥ 10 weeks, ≥ 1 abortion	8 (20) 6 (15)	3 (15) 1 (5)	5 (25) 5 (25)	0.34 0.09
• Pre-eclampsia/eclampsia • <34 weeks, ≥ 1 premature birth				
Livedo reticularis	11 (11.2)	2 (4.8)	9 (16.1)	0.07
Thrombocytopenia	33 (33.7)	9 (21.4)	24 (42.9)	**0.02**
APS nephropathy	9 (9.2)	2 (4.8)	7 (12.5)	0.17
Valvular heart disease	33 (33.7)	12 (28.6)	21 (37.5)	0.24
Conventional risk factors	53 (53.1)	17 (40.5)	35 (62.5)	
• Arterial hypertension	47 (48)	21 (50)	26 (46.4)	**0.02**
• Hyperlipidemia	6 (6.1)	3 (7.1)	3 (5.4)	0.41
• Diabetes mellitus	35 (35.7)	19 (45.2)	16 (28.6)	0.51
• Obesity	30 (30.6)	12 (28.6)	18 (32.1)	0.07
• Smoking				0.43
aPL profile	77 (78.6)	29 (69)	48 (85.7)	
• LA	59 (60.2)	31 (73.8)	28 (52.8)	**0.04**
• aCL IgG/IgM	49 (50)	22 (56.4)	27 (52.9)	**0.03**
• aβ2GPI IgG/IgM	30 (30.6)	14 (33.3)	17 (30.4)	0.45
• Triple positive				0.46

Methods Ninety-eight patients with APS were classified according to clinical manifestations as vascular thrombosis (VT), pregnancy morbidity (PM) or both (VT+PM). The aGAPSS was calculated as defined before. Arterial hypertension and hyperlipidemia definitions were made according to the ESC/ESH ve NCEP/ATP III guidelines, respectively.

Results Demographic, laboratory and clinical characteristics of patients are summarized in table 1. Mean aGAPSS was calculated as 10.2 ± 3.8. Significantly higher aGAPSS values were seen in VT (n=58) and VT+PM (n=29) compared to PM (n=11) (mean aGAPSS 10.6 ± 3.7 vs 7.3 ± 2.9, P=0.005; 10.5 ± 4 vs 7.3 ± 2.9, P=0.01, respectively). AUC demonstrated that aGAPSS values ≥ 10 had the best diagnostic accuracy for thrombosis (AUC: 0.71, sensitivity: 0.52, specificity: 0.91, P=0.01). Higher aGAPPS values were also associated with recurrent thrombosis (mean aGAPSS 11.5 ± 3.7 vs 9.9 ± 3.6, P=0.04). Regarding extra-criteria manifestations, patients with livedo reticularis (n=11) and APS nephropathy (n=9) had significantly higher aGAPSS values (mean aGAPSS 12.9 ± 3.4 vs 9.9 ± 3.7, P=0.02; 12.4 ± 2.9 vs 10 ± 3.8, P=0.04, respectively).

Conclusion Our results suggest that patients with higher aGAPSS values are at higher risk for developing vascular thrombosis (either single or recurrent) and extra-criteria manifestations, especially livedo reticularis and APS nephropathy.

P10 ANTIPHOSPHOLIPID ANTIBODIES ASSOCIATED EXTRA-CRITERIA CLINICAL MANIFESTATIONS SHOULD NOT BE IGNORED

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Background The current Sydney classification criteria do not consider a range of non-thrombotic clinical manifestations that are frequently observed in association with the presence of aPLs, the so-called extra-criteria manifestations. The aim of this study was to retrospectively analysis our single center Antiphospholipid antibodies associated clinical phenotypes, especially the frequency of extra-criteria manifestations.

Methods Data of 731 serum samples from patients of clinical suspected APS in 2018 were enrolled. Data of clinical features, laboratory examination, treatment and prognosis were retrospectively analyzed.

Results A total of 200 patients with APLs were positive (27%), 56 males and 144 females, with an average age of 40.13±17.24 years, 115 cases (57.5%) with ACL positive, 167 cases (83.5%) with anti-β2GPI antibodies positive, 69 cases (34.5%) with LA positive, and 20 cases (10%) turned negative after 12 weeks without any specific therapy. 61 (30.5%) patients were fulfilled the 2006 revised Sydney classification criteria for APS. One patient was Catastrophic APS. 27(44.3%) patients with primary APS, 34 (55.6%) were secondary APS, including SLE-APS (20), CTD (9), infection (3), malignancies (2). 119 patients with persistent APLs positive, but were not fulfilled the APS criteria because of without thrombosis or fetal loss. 77 (42.8%) patients were asymptomatic, 42 patients with extra-criteria manifestations. The total frequency of extra-criteria manifestations associated with APLs are 40%. The details are shown in table 1.

Abstract P10 Table 1 Extra-criteria clinical manifestations of APS and APLs-carriers

	PAPS (N=27)	Secondary APS(N=34) SLE (N=20)	CTD (N=9)	Non-AID (N=5)	Total	APLs persist positive(N=119) SLE (N=40)	CTD (N=26)	Non-AID (N=15)	Unknown (N=38)	Total	Total (N=180)
Hematological	6	11	5	2	18	19	2	4	8	33	57 (31.7%)
Thrombocytopenia	6	10	4	1	15	18	2	4	8	32	53 (29.4%)
Haemolytic anaemia	1	3	2	2	7	-6	0	0	0	6	14 (7.8%)
Livedo reticularis	0	1	1	0	2	1	0	0	0	1	3 (1.7%)
APS nephropathy	2	1	2	1	4	0	0	0	0	0	6 (3.3%)
Valvular heart disease	2	6	1	0	7	4	0	1	1	6	15 (8.3%)
Non-vascular neurological manifestations	3	3	4	1	8	4	1	0	2	7	18 (10%)
Thrombophlebitis	0	2	0	1	3	0	2	1	0	3	6 (3.3%)
Total	**9**	**13**	**6**	**2**	**21**	**21**	**5**	**5**	**11**	**42**	**72 (40%)**

Conclusions The most common extra-criteria manifestations are thrombocytopenia, non-vascular neurological manifestations and valvular heart disease. And they can be the independent clinical feature of APLs without thrombotic events or Pregnancy morbidities.

P11 EARLY EFFICIENT ANTICOAGULATION IMPROVES THE LONG-TERM PROGNOSIS IN PATIENTS WITH ANTIPHOSPHOLIPID SYNDROME ASSOCIATED PORTAL VEIN THROMBOSIS

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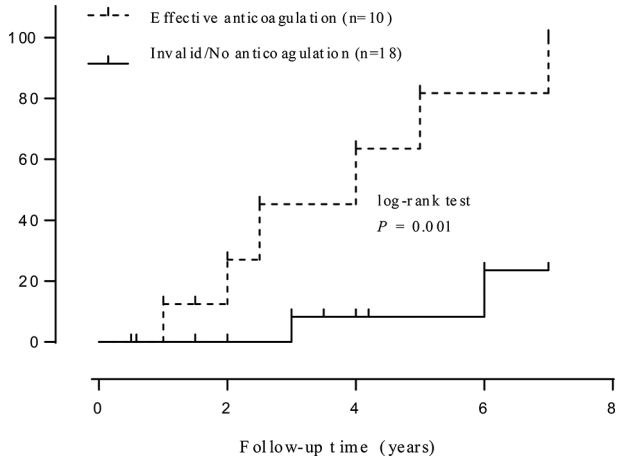
Background Portal vein thrombosis (PVT) is a rare and severe clinical phenotype of antiphospholipid syndrome (APS) with a poor prognosis. Anticoagulation therapy is efficient, but is associated with potentially severe side-effects, especially bleeding episodes. The aim of this study was to retrospectively analyze our single center experience on long-term anticoagulation in APS patients presenting a PVT.

Methods A retrospective study of APS patients with PVT from 2012 to 2019 was conducted using the Hospital Information System of Peking Union Medical College Hospital. Basic clinical history and complications were collected. Regular imaging was performed to monitor the outcome of PVT. The recanalization rate of the PVT after anticoagulation was analyzed using the survival analysis.

Results A total of 28 patients with APS-PVT were enrolled, 5 males and 23 females, with the median age 37 years (range 17–63 years), and the mean follow-up was 3 years (range, 0.5–7 years). 8 cases were acute thrombosis, 16 cases chronic thrombosis, and 4 cases portal vein cavernoma. The first symptoms presented as abdominal distention (14/28) or pain (7/28) and blood system involvements (22/28, anemia or thrombocytopenia), while presentation with variceal bleeding (4 cases) was less common, and 2 patients were asymptomatic. Triple aPLs positive in 7 cases. 10 cases began efficient anticoagulation therapy immediately at the diagnosis of thrombus. 8 patients got thrombus recanalization. 3 patients got recurrence. 5 patients died. Survival analysis revealed that effective anticoagulation could increase recanalization rate significantly (log rank p =0.001), as shown in figure 1.



Abstract P11 Figure 1 Difference of accumulated recanalization rate between receiving immediate effective anticoagulation group and invalid anticoagulation group

Conclusions PVT usually had insidious onset with atypical clinical symptoms and easily be misdiagnosed. Early diagnosis and efficient anticoagulation treatment can bring thrombus recanalization thereby significantly improving the prognosis.

P12 SERUM LEVELS OF SOLUBLE ST2 AND THEIR ASSOCIATION WITH MICROPARTICLES AND DISEASE ACTIVITY IN SYSTEMIC LUPUS ERYTHEMATOSUS

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Background ST2 is an IL-33 receptor (a member of the IL-1 receptor family), existing in a transmembrane form (ST2L) and is also alternatively spliced to produce a secreted soluble form (sST2) and a membrane-anchored variant without the immunoglobulin-like motif (ST2V). High levels of sST2 have been reported in inflammatory diseases, including systemic lupus erythematosus (SLE). Additionally, higher levels of