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

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**Background/Purpose** Adjusted global antiphospholipid 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)	5 (25)	4 (20)	0.5
• ≥ 10 weeks, ≥ 1 abortion	29 (72.5)	14 (70)	15 (75)	0.5
• ≥ 10 weeks, ≥ 1 abortion	8 (20)	3 (15)	5 (25)	0.34
• Pre-eclampsia/eclampsia	6 (15)	1 (5)	5 (25)	0.09
• <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  $10.2 \pm 3.8$ . Significantly higher aGAPSS values were seen in VT ( $n=58$ ) and VT+PM ( $n=29$ ) compared to PM ( $n=11$ ) (mean aGAPSS  $10.6 \pm 3.7$  vs  $7.3 \pm 2.9$ ,  $P=0.005$ ;  $10.5 \pm 4$  vs  $7.3 \pm 2.9$ ,  $P=0.01$ , respectively). AUC demonstrated that aGAPSS values  $\geq 10$  had the best diagnostic accuracy for thrombosis (AUC: 0.71, sensitivity: 0.52, specificity: 0.91,  $P=0.01$ ). Higher aGAPSS values were also associated with recurrent thrombosis (mean aGAPSS  $11.5 \pm 3.7$  vs  $9.9 \pm 3.6$ ,  $P=0.04$ ). Regarding extra-criteria manifestations, patients with livedo reticularis ( $n=11$ ) and APS nephropathy ( $n=9$ ) had significantly higher aGAPSS values (mean aGAPSS  $12.9 \pm 3.4$  vs  $9.9 \pm 3.7$ ,  $P=0.02$ ;  $12.4 \pm 2.9$  vs  $10 \pm 3.8$ ,  $P=0.04$ , 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.

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### 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  $40.13 \pm 17.24$  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.