Results: We were able to examine 91 AAV patients (52 MPA patients and 39 GPA patients) with 82.4% for MPO-ANCA positive and 20.9% for PR3-ANCA positive. Almost half of the patients was female (56.0%). The median age was 70 years [interquartile range (IQR): 64-77]. The median BVAS was 17 (IQR: 12-23). We identified autoantigen of EGF-containing fibulin-like extracellular matrix protein 1 (EFEMP1) in 43 of MPA (82.6%) and 16 of GPA (41.0%) at baseline. After 6 months of treatment, no cases of EFEMP1 were identified in MPA and GPA. The clinical features of EFEMP1 positive in AAV patients were higher age at onset (p <0.01), less ear, nose and throat symptoms at initiation of treatment (p <0.05), higher serum Cr at initiation of treatment (p <0.01), higher vasculitis damage index (VDI) renal component at 12 months and 24 months after initiation of treatment (both p <0.05).

Conclusion: Our findings indicate that an autoantigen as immune complexes of EFEMP1 were involved in the pathogenesis of AAV patients and may predict renal prognosis.

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Table. Comparison with and without EFEMP1 (all cases)

group	EFEMP1 positive (n=59)	EFEMP1 negative (n=32)	**p-value
Sex(%male)	27/59 (45.8%)	13/32 (40.6%)	0.665
Age, years	74 (66-78)	68 (60-71)	0.003
WBC(/ml)	8270 (7325-12725)	9150 (7325-11700)	0.280
Cr(mg/dl)	1.4 (0.9-3.8)	0.8 (0.6-1.5)	0.005
CRP(mg/dl)	7.0 (2.3-12.5)	7.6 (4.0-11.0)	0.566
MPA(%)	43/59 (72.9%)	9/32 (28.1%)	<0.001
GPA(%)	16/59 (27.1%)	23/32 (71.9%)	<0.001
MPO-ANCA positive	54/59 (91.5%)	24/32 (65.6%)	0.003
PR3-ANCA positive*	7/56 (12.5%)	12/32 (37.5%)	0.012
BVAS total	15 (12-20)	20 (12-25)	0.087
BVAS renal positive	53/59 (89.8%)	26/32 (81.3%)	0.332
BVAS chest positive	16/59 (27.1%)	14/32 (43.4%)	0.161
BVAS ENT positive	17/59 (28.8%)	18/32 (56.3%)	0.014
BVAS systemic positive	40/59 (67.8%)	24/32 (75.0%)	0.631
VDI renal 6 months	1 (0-2)	0 (0-1)	0.053
VDI renal 12 months	1 (0-2)	0 (0-1)	0.007
VDI renal 24 months	1 (0-2)	0 (0-1)	0.012

IQR interquartile range. Values are median(IQR) or n(%), *missing data, **Wilcoxon signed-rank test/Fisher's exact test

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THU0316 VALIDATION OF ACR/EULAR PROVISIONAL CLASSIFICATION CRITERIA FOR ANCA-ASSOCIATED VASCULITIS IN A LATIN-AMERICAN TERTIARY CENTER

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Background: There is lack studies about performance of new criteria set for ANCA-Associated Vasculitis (AAV) in Latin-America.

Objectives: To validate the new classification criteria for AAV in a real-life cohort of patients with these conditions.

Methods: We performed a review of medical records from January 1990 to December 2019 at Hospital Nacional Guillermo Almenara Irigoyen from Peru. AAV was diagnosed by experienced rheumatologists based on the ACR 1990 criteria, Chapel Hill 2012 consensus, EMEA criteria and their experience and clinical acumen. Granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA) were diagnosed. Renal limited vasculitis was considered as MPA. To evaluate the performance of the new criteria, we classified all patients using "former criteria set" (including the 1990 ACR criteria for GPA and EGPA and the 1994 Chapel Hill Consensus Conference for MPA) and the EMEA (European Medicines Agency) criteria set. At the same time, we classified all patients using the ACR/EULAR Provisional criteria (new criteria set). The values for sensitivity, specificity and level of agreement (using Cohen's kappa) of all sets of criteria were calculated using the clinical diagnosis as gold standard.

Results: Two hundred twelve patients were identified; 12 of them were excluded (eight did not have ANCA and four had incomplete data). Female/male ratio was 1.9:1 [130 (65%)/70 (35%)] and their mean (SD) age at diagnosis was 59.3 (12.6) years. One hundred fifty-four (77%) had MPA, 41 (20.5%) GPA and 5 (2.5%) EGPA. One hundred ninety-six patients had ANCA-IIF results [p-ANCA: 131 (66.8%), c-ANCA: 43 (21.9%), negative-ANCA: 22 (11.3%)] and 190 patients had ANCA-ELISA results [MPO: 129 (67.9%), PR3: 37 (19.5%), negative-ANCA: 24 (12.6%)]. Type of diagnosis according to criteria set used is depicted in Table 1. The new criteria set had better agreement (kappa: 0.653) than the EMEA criteria (kappa: 0.506) and the former criteria set (kappa: 0.305). Performance of the criteria sets is depicted in Table 2.

Table 1. Type of AAV according to criteria set used.

TYPE OF AAV	Clinical diagnosis	Former criteria	New criteria	EMEA criteria
MPA, n (%)	154 (77)	76 (38)	137 (68.5)	110 (56.0)
GPA, n (%)	41 (20.5)	30 (15)	39 (19.5)	39 (19.5)
EGPA, n (%)	5 (2.5)	2 (1)	4 (2)	2 (1.0)
Not classifiable, n (%)	NA	92 (46)	20 (10)	44 (22.0)
PAN				5 (2.5)

PAN: Polyarteritis nodosa. NA: Not applicable.

Table 2. Performance of the different criteria sets in AAV patients.

DIAGNOSIS	CRITERIA SET	SE	SP	Kappa
MPA	Former	49.4	100.0	0.309
	EMEA	69.9	93.9	0.471
	New	87.0	93.5	0.713
GPA	Former	68.3	98.7	0.744
	EMEA	92.7	99.4	0.938
	New	80.5	96.2	0.781
EGPA	Former	40.0	100.0	0.565
	EMEA	40.0	100.0	0.565
	New	60.0	99.5	0.659

SE: Sensitivity. SP: Specificity.

Conclusion: The ACR/EULAR Provisional Criteria for AAV have better agreement with the clinical diagnosis of AAV in Latin-American patients from a real-life cohort.

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THU0317 EXPANDED DOUBLE NEGATIVE T CELLS IN PATIENTS WITH ANTINEUTROPHIL CYTOPLASMIC AUTOANTIBODY ASSOCIATED VASCULITIS PRODUCE CYTOKINES AND INDUCE RENAL DAMAGE

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