diagnosis or exclusion of GCA were done on median 1 day (in 55% of patients). However, in cases where further diagnostics (TAB and PET-CT) were necessary, the time for diagnosis increased substantially. See table 2 for data on waiting time from referral, diagnoses and time required for diagnosis.

Table 1

Demographic and clinical data (n=120)	
Females	72%
Age (years)	74 (46-98)
Symptom duration (days)	16 (1-180)
New onset headache	72%
Jaw claudication	21%
Temporal artery abnormality	33%
Visual symptoms	42%
Visual loss (permanent visual loss)	18% (19%)
Constitutional symptoms	32%
Fever	11%
Weight loss	18%
Polymyalgia symptoms	33%
Glucocorticoid therapy	68%
Duration of glucocorticoid (days)	3 (1-365)
Haemoglobin (mmol/L)	8.0 (5.0-10.0)
C-reactive protein (mg/L)	22 (0-290)
Sedimentation rate (mm/hr)	38 (2-146)

Values are percentage, median (range)

Table 2

Office days from referral to visit	1 (0-20)
Days for diagnosis/exclusion of GCA	1 (1-46)
GCA clinical diagnosis	35%
GCA ACR1990 criteria	42%
Nr. of US (% w. GCA)	120 (36%)
Nr. of PET-CT (% w. GCA)	44 (23%)
Nr. of TAB (% w. GCA)	34 (21%)
Polymyalgia diagnosis	17%
Arthritis diagnosis	5%
Other diagnosis	17%

Conclusion: In this newly established GCA FTC, fast and precise diagnosis of GCA or exclusion of this was possible in majority of patients referred, even though referral criteria were liberal. In 55% of patients, further diagnostic procedures were considered unnecessary, and a diagnosis on day 1 could be established based on US, clinical findings and existing paraclinical data.

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THU0314 RELAPSES AND LONG-TERM REMISSION IN LARGE VESSEL GIANT CELL ARTERITIS IN NORTHERN ITALY: CHARACTERISTICS AND PREDICTORS IN A LONG-TERM FOLLOW-UP STUDY

L. Boiardi¹, E. Muratore¹, G. Restuccia¹, A. Cavazza², M. G. Catanoso¹, P. Macchioni¹, L. Spaggiari³, L. Cimino⁴, R. Aldigeri⁵, N. Pipitone¹, A. Fontana⁶, M. Csaali⁷, S. Croci⁸, N. Girolimetto¹, C. Salvarani⁹.

¹Rheumatology Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy;

²Pathology Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy;

³Radiology Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy;

⁴Ophthalmology Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy;

⁵University of Parma, Parma, Italy; ⁶Vascular Surgery Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy; ⁷Nuclear Medicine Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy; ⁸AABI, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy; ⁹Radiology Unit, AUSL-IRCCS di Reggio Emilia and University of Modena and Reggio Emilia, Reggio Emilia, Italy

Background: Previous studies evaluated clinical relapses and long-term remission mainly in patients with biopsy-proven GCA and/or patients satisfying the ACR 1990 criteria for GCA classification. Also, radiological involvement was unfrequently used to define relapses and monitor disease activity in patients with LV-GCA

Objectives: To evaluate characteristics and predictors of relapses and long-term remission in an Italian cohort of patients with large-vessel (LV) giant cell arteritis (GCA).

Methods: We evaluated 87 consecutive patients with LV-GCA followed up at the Rheumatology Unit of Reggio Emilia Hospital (Italy) for at least 2 years. Patients with relapses and long-term remission were compared to those without. A comparison group of 34 patients with biopsy proven GCA without LV vasculitis (LVV) at diagnosis was considered for comparison.

Results: 37 patients (42.5%) experienced one or more relapses. Nineteen (37.2%) of the 51 relapses were experienced during the first year after diagnosis. The majority of relapses occurred with doses of prednisone (PDN) $\leq$ 10 mg/day (74.5%). Polymyalgia rheumatica (PMR) (41.2%) and worsening at imaging of LVV (39.2%) were the most frequently observed relapsing manifestations. The total cumulative prednisone dose was significantly higher ($p = 0.0001$) and the total duration of PDN treatment longer ($p = 0.0001$) in relapsing patients compared to those without relapses. Relapsing patients had at diagnosis more frequently fever $\geq 38^\circ\text{C}$ ($p = 0.03$) and visual manifestations ($p = 0.03$), and less frequently long-term remission ($p = 0.003$). In the multivariate model fever $\geq 38^\circ\text{C}$ (HR 3.22, 95%CI: 1.43-7.27), duration of PDN treatment (HR 1.01, 95%CI: 1.00-1.02) and total cumulative PDN dose (HR 1.20, 95%CI: 1.09-1.33) were significantly associated with an increased risk of relapses, while aortic arch involvement at imaging at diagnosis (HR 0.22, 95%CI: 0.09-0.55) and long-term remission (HR 0.30, 95%CI: 0.12-0.75) with a reduced risk. 37 patients (42.5%) experienced long-term remission. PMR and disease relapses were less frequently observed ($p = 0.003$ for both), and the total cumulative prednisone dose was lower ($p = 0.005$) in patients with long-term remission compared to those without. In the multivariate model the presence of relapses (HR 0.33, 95%CI: 0.14-0.78) and the total cumulative PDN dose (HR 0.89, 95%CI: 0.83-0.96) were significantly negatively associated with long-term remission.

Conclusion: In our cohort of patients with LV GCA we identified predictors of a relapsing course and long-term remission, which were observed in around half of the patients.

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THU0315 THE CLINICAL RELEVANCE OF SERUM IMMUNE COMPLEXES IN ANCA-ASSOCIATED VASCULITIS

M. Okamoto¹, K. Ichinose¹, K. Oyama², K. E. Sada³, S. Y. Kaname⁴, M. Harigai⁵, A. Kawakami¹. ¹Nagasaki University Graduate School of Biomedical Sciences, Division of Advanced Preventive Medical Sciences, Department of Immunology and Rheumatology, Nagasaki, Japan; ²Nagasaki University Graduate School of Biomedical Sciences, Department of Pharmacy Practice, Nagasaki, Japan; ³Okayama University Graduate School of Biomedical Sciences, Department of Nephrology, Rheumatology, Endocrinology and Metabolism, Okayama, Japan; ⁴Kyorin University School of Medicine, Department of Nephrology and Rheumatology, Tokyo, Japan; ⁵Tokyo Women's Medical University, Department of Rheumatology, Tokyo, Japan

Background: AAV is an autoimmune disease characterized by systemic vasculitis and pauci-immune-type crescentic glomerulonephritis (CGN) with ANCA production¹. Several authors have reported cases of ANCA-associated CGN with definite IC deposits^{2,3}, however, the clinical significance of IC in patients with ANCA-associated CGN remains unclear.

Objectives: To investigate the clinical relevance of serum immune complexes (ICs) in ANCA-associated vasculitis (AAV) patients.

Methods: We developed a novel proteomic strategy for identifying and profiling antigens in immune complexes in the serum of microscopic polyangiitis (MPA) and granulomatosis with polyangiitis (GPA) patients. The serum was collected from the cohort of Japan Research Committee of the Ministry of Health Labour, Welfare for Intractable Vasculitis (JPVAS) before treatment and 6 months after initiation of treatment. The serum from healthy individuals was used for control samples. The Baseline data of each patient was collected with demographic information, laboratory data, disease activity according to the Birmingham Vasculitis Activity Score (BVAS) 2003, disease severity, and imaging data.

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

V. Pimentel-Quiroz^{1,2}, A. Sánchez-Torres¹, C. Reategui Sokolova^{1,3}, R. V. Gamboa Cárdenas¹, C. Sánchez-Schwartz¹, M. Medina Chinchon¹, F. Zevallos Miranda¹, E. Noriega¹, J. Alfaro Lozano¹, J. M. Cucho-V¹, Z. Rodríguez Bellido^{1,4}, C. Pastor Asurza^{1,4}, E. Acevedo-Vásquez^{1,4}, R. Perich Campos^{1,4}, G. S. Alarcon^{5,6}, M. F. Ugarte-Gil^{1,2}. ¹Hospital Nacional Guillermo Almenara Irigoyen, Rheumatology, Lima, Peru; ²Universidad Científica del Sur, Lima, Peru; ³Universidad San Ignacio de Loyola, Unidad de Investigación para la Generación y Síntesis de Evidencias en Salud, Lima, Peru; ⁴Universidad Nacional Mayor de San Marcos, Lima, Peru; ⁵University of Alabama at Birmingham, Division of Clinical Immunology and Rheumatology, Department of Medicine, School of Medicine, Birmingham, United States of America; ⁶Universidad Peruana Cayetano Heredia, Medicina, Lima, Peru

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

Y. Qin¹, J. Luo², Y. Wang¹, C. Gao³. ¹The Second Hospital of Shanxi Medical University, Taiyuan, China; ²The Second Hospital of Shanxi Medical University,