2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis

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ABSTRACT

Objective To develop and validate updated classification criteria for giant cell arteritis (GCA).

Methods Patients with vasculitis or comparator diseases were recruited into an international cohort. The study proceeded in six phases: (1) identification of candidate items, (2) prospective collection of candidate items present at the time of diagnosis, (3) expert panel review of cases, (4) data-driven reduction of candidate items, (5) derivation of a points-based risk classification score in a development data set and (6) validation in an independent data set.

Results The development data set consisted of 518 cases of GCA and 536 comparators. The validation data set consisted of 238 cases of GCA and 213 comparators. Age ≥50 years at diagnosis was an absolute requirement for classification. The final criteria items and weights were as follows: positive temporal artery biopsy or temporal artery halo sign on ultrasound (+5); erythrocyte sedimentation rate ≥50 mm/hour or C reactive protein ≥10 mg/L (+3); sudden visual loss (+3); morning stiffness in shoulders or neck, jaw or tongue claudication, new temporal headache, scalp tenderness, temporal artery abnormality on vascular examination, bilateral axillary involvement on imaging and fluorodeoxyglucose–positron emission tomography activity throughout the aorta (+2 each). A patient could be classified as having GCA with a cumulative score of ≥6 points. When these criteria were tested in the validation data set, the model area under the curve was 0.91 (95% CI 0.88 to 0.94) with a sensitivity of 87.0% (95% CI 82.0% to 91.0%) and specificity of 94.8% (95% CI 91.0% to 97.4%).

Conclusion The 2022 American College of Rheumatology/EULAR GCA classification criteria are now validated for use in clinical research.

INTRODUCTION

Giant cell arteritis (GCA), formerly known as temporal arteritis, is the most common form of systemic vasculitis in patients aged ≥50 years.[1] GCA is defined by granulomatous arteritis that affects large-sized and medium-sized blood vessels with a predisposition to affect the cranial arteries.[2] Common presenting features of the disease include headache, constitutional symptoms, jaw claudication, scalp tenderness, visual disturbances and elevated inflammatory markers.[3]

In 1990, the American College of Rheumatology (ACR) endorsed classification criteria for GCA.[4] These criteria were established before the widespread use of non-invasive and advanced vascular imaging modalities, which have become increasingly incorporated in the clinical assessment of GCA. Vascular ultrasound can be used to diagnose GCA, and depending on the clinical setting, a non-compressible 'halo' sign of a temporal±axillary artery may replace the need for temporal artery biopsy (TAB).[5–8] Moreover, vascular imaging has demonstrated that arterial involvement in GCA is not exclusively confined to the cranial arteries[9 10] and can commonly affect the aorta and primary branches in a pattern similar to Takayasu arteritis (TAK).[11 12]

The limitations of the ACR 1990 criteria for GCA have become more apparent in the conduct of recent clinical trials and other research studies, in which investigators typically modify the 1990 ACR criteria to reflect modern practice.[6 13 14] Notably, the 1990 ACR criteria focus mostly on cranial features of GCA and do not perform well in classifying patients with disease predominantly affecting the larger arteries. The 1990 ACR criteria were derived by using comparator populations, which included many patients with small-vessel vasculitis, a form of vasculitis that is not typically difficult to differentiate from GCA. In addition, the 1990 ACR criteria for GCA followed the 'number of criteria' rule, which considered each criterion to have equal weight as a classifier for the disease. Since then, methodologic advances in classification criteria have allowed movement towards weighted criteria with threshold scores that improve performance characteristics.[15 16]

This article outlines the development and validation of the revised ACR/EULAR-endorsed classification criteria for GCA.

METHODS

An international Steering Committee comprising clinician investigators with expertise in vasculitis, statisticians and data managers was assembled to oversee the overall development of classification criteria for primary vasculitis.[17] A detailed and complete description of the methods involved in the development and validation of the classification criteria for GCA is located in online supplemental appendix 1. Briefly, the Steering Committee implemented a six-stage plan using data-driven and consensus methodology to develop the following criteria.

Stage 1: generation of candidate classification items for the systemic vasculitides

Candidate classification items were generated by expert opinion and reviewed by a group of vasculitis experts across a range of specialties using nominal group technique.

Stage 2: Diagnostic and Classification Criteria for Vasculitis prospective observational study

A prospective, international, multisite observational study was conducted (see online supplemental file 3 for study investigators and sites). Consecutive patients representing the full spectrum of vasculitides were recruited from academic and community practices. Patients were included if they were 18 years or older and had a diagnosis of vasculitis or a condition that mimics vasculitis (eg, infection, malignancy, atherosclerosis). Patients with GCA could only be enrolled within 2 years of diagnosis. Only data present at diagnosis were used to develop the classification criteria.

Stage 3: expert review to derive a gold standard-defined set of cases of large-vessel vasculitis

Experts in vasculitis from a wide range of geographic locations and specialties (see online supplemental file 3) reviewed all submitted cases of vasculitis and a random selection of vasculitis mimics. Each reviewer was asked to review ~50 submitted cases to confirm the diagnosis and to specify the degree of certainty of their diagnosis as follows: very certain, moderately certain, uncertain or very uncertain. Only cases agreed on with at least moderate certainty by two reviewers were retained for further analysis.

Stage 4: refinement of candidate items specifically for large-vessel vasculitis

The Steering Committee conducted a data-driven process to reduce the number of candidate items of relevance to cases and comparators for large-vessel vasculitis (LVV). Density plots were assessed to study age distribution at diagnosis and symptom onset for GCA and TAK. Absolute age requirements vs incorporation of age as a candidate criteria item was considered. Items related to the vascular physical examination, vascular imaging, arterial biopsy and laboratory values were combined or eliminated based on consensus review. Items were selected for exclusion if they had a prevalence of <5% within the data set, and/or they were not clinically relevant for classification criteria (eg, related to infection, malignancy or demography). Low-frequency items of clinical importance could be combined, when appropriate. Patterns of vascular imaging findings detected by vascular ultrasound, angiography or positron emission tomography (PET) were defined by K-means clustering.[18]

Stage 5: derivation of the final classification criteria for GCA

The Diagnostic and Classification Criteria for Vasculitis (DCVAS) data set was split into development (70%) and validation (30%) sets. Comparisons were performed between cases of GCA and a randomly selected comparator group in the following proportions: TAK, 33.5%; other vasculitides that mimic GCA and TAK (isolated aortitis, primary central nervous system vasculitis, polyarteritis nodosa, Behçet's disease and other LVV), 33.4%; and other diagnoses that mimic LVV (eg, atherosclerosis, unspecific headache), 33.1%. Least absolute shrinkage and selection operator (lasso) logistic regression was used to identify predictors from the data set and create a parsimonious model including only the most important predictors.[19] The final items in the model were formulated into a clinical risk-scoring tool, with each factor assigned a weight based on its respective regression coefficient. A threshold that best balanced sensitivity and specificity was identified for classification.

Stage 6: validation of the final classification criteria for GCA

Performance of the new criteria was validated in an independent set of cases and comparators. Performance of the final classification criteria was examined in specific subsets of patients with GCA using data from the combined development and validation sets to maximise sample sizes for the subgroups. Patients were studied according to different disease subtypes (biopsy-proven GCA and large-vessel GCA) and regions of the world (North America, Europe) where clinical strategies to assess GCA are known to differ.[20] Biopsy-proven GCA was defined as definite vasculitis on TAB reported by the submitting physician, and large-vessel GCA was defined as vasculitic involvement of the aorta or its branch arteries on either angiography (computed tomography, magnetic resonance imaging, or catheter-based angiography), ultrasound or PET, without vasculitis on TAB. Comparison was made between the measurement properties of the new classification criteria for GCA and the 1990 ACR classification criteria in the validation data set. Performance characteristics of the criteria were also tested in patients with TAK and compared with those with GCA diagnosed between the ages of 50 and 60 years.

RESULTS

Generation of candidate classification items for the systemic vasculitides

The Steering Committee identified >1000 candidate items for the DCVAS Case Report Form (see online supplemental appendix 2).

DCVAS prospective observational study

Between January 2011 and December 2017, the DCVAS study recruited 6991 participants from 136 sites in 32 countries. Information on the DCVAS sites, investigators and study participants is listed in online supplemental appendices 3, 4 and 5.

Expert review methodology to derive a gold standard-defined final set of cases of LVV

The LVV expert panel review process included 56 experts who reviewed vignettes derived from the Case Report Forms for 2131 cases submitted with a diagnosis of LVV (1608 (75.5% of Case Report Forms)), another type of vasculitis (118 (5.5% of Case Report Forms)) or a mimic of vasculitis (405 (19.0% of Case Report Forms)). Characteristics and the list of expert reviewers are shown in online supplemental appendices 6 and 7. A sample vignette and the LVV expert panel review flow chart are shown in online supplemental appendices 8 and 9. A total of 1695 cases (80%) passed the main LVV process. An additional 373 cases of LVV and comparators, confirmed during a previous review process to derive the classification criteria for antineutrophil cytoplasmic antibody-associated vasculitis, were also included. In total, after both review processes, 2068 cases were available for the stages 4 and 5 analyses.

The submitting physician diagnosis of GCA was confirmed in 913 of 1137 cases (80.3%) after both expert panel reviews. The reasons for exclusion were diagnosis of GCA categorised as 'uncertain' or 'very uncertain' during panel review (n=187) or change in diagnosis during panel review to another type of vasculitis (isolated aortitis, TAK, other vasculitides) (n=11) or

to a comparator disease (n=26). An additional 29 patients who were not initially diagnosed as having GCA by the submitting physician were diagnosed as having GCA after panel review and DCVAS Steering Committee member adjudication. In total, 942 cases of confirmed GCA were available for analysis. To balance the number of cases of GCA with the number of available comparators, 756 cases of GCA were randomly selected for subsequent analysis.

Refinement of candidate items specifically for GCA

Only 7 of 942 patients with GCA (<1%) were diagnosed at age <50 years (see online supplemental appendix 10 for the distribution of 'age at diagnosis' in patients with LVV, and the similar distribution of 'age at symptom onset,'). Therefore, an age of ≥50 years at diagnosis was considered an absolute requirement to classify GCA. Cluster analyses of vascular imaging data identified bilateral axillary involvement and diffuse fluorodeoxyglucose uptake throughout the aorta on PET as specific imaging patterns for GCA (see online supplemental appendices 11 and 12). These imaging patterns were tested as potential classifiers.

Following a data-driven and expert consensus process, 72 items of the DCVAS Case Report Form were retained for regression analysis, including 32 demographic and clinical items, 14 laboratory items (including values of C reactive protein (CRP) level and erythrocyte sedimentation rate (ESR), each divided into 5 categories), 14 imaging items (13 composite), 11 vascular examination items (5 composite and upper extremity blood pressure divided into 6 categories) and 1 biopsy item (online supplemental appendix 13).

Derivation of the final classification criteria for GCA

A total of 1505 patients were selected for analysis (756 GCA and 749 comparators), of which 1054 (70%) were in the development data set (518 GCA and 536 comparators) and 451 (30%) in the validation data set (238 GCA and 213 comparators). Table 1 describes the demographic and clinical features of the patients with GCA and the comparators. The patients with GCA were recruited from Europe (n=796), North America (n=112), Oceania (n=18) and Asia (n=16). Clinical diagnoses assigned to patients in the comparator group are detailed in online supplemental appendix 14.

Lasso regression of the previously selected 72 items yielded 27 independent predictor variables for GCA (online supplemental appendix 15A). Each predictor variable was then reviewed for inclusion by the DCVAS Steering Committee, based on their ORs and specificity to GCA, to ensure face validity. The variables 'definitive vasculitis on TAB' and 'halo sign on temporal artery ultrasound' were found to dominate the model as quite strong predictors of GCA (see online supplemental appendix 16A for cluster plots showing almost a perfect overlap between the diagnosis of GCA and positive TAB or halo sign on temporal artery ultrasound). Therefore, for the remaining variables to have discriminatory value, both of these items were removed from the model, combined into one composite item 'vasculitis on TAB or halo sign on temporal artery ultrasound' and given a risk score of one point below the final threshold set to classify GCA to maintain face validity. The variables 'jaw claudication' and 'tongue claudication' were combined into one item, as were the variables 'maximum ESR (>50 mm/hour)' and 'maximum CRP (>10 mg/L).' Although the variable 'new persistent headache, occipital or cervical' showed important statistical significance, it decreased the overall specificity of the model when testing their final performance characteristics (patients vs comparators) and was, therefore, also removed. Weighting of the individual criterion included in the model was based on logistic regression fitted to the remaining nine selected predictors (online supplemental appendix 17A).

Table 1 Demographic and disease features of the patients with giant cell arteritis and the comparators*

	GCA (n=756)	Comparators (n=749)†	P value
Age, mean±SD years	72.2±8.5	44.6±18.0	<0.001
Female sex	511 (67.6)	447 (59.7)	0.001
Clinical features			
Morning stiffness, neck/torso	88 (11.6)	15 (2.0)	<0.001
Morning stiffness, shoulders/arms	174 (23.0)	23 (3.1)	<0.001
Sudden visual loss	102 (13.5)	29 (3.9)	<0.001
Jaw claudication	356 (47.1)	19 (2.5)	<0.001
Tongue claudication	21 (2.8)	1 (0.1)	<0.001
New persistent temporal headache	475 (62.8)	90 (12.0)	<0.001
Scalp tenderness	260 (34.4)	25 (3.3)	<0.001
Temporal artery abnormality on vascular examination‡	354 (46.8)	35 (4.7)	<0.001
Investigations			
Maximum ESR ≥50 mm/hour	558 (73.8)	291 (38.9)	<0.001
Maximum CRP ≥10 mg/L	683 (90.3)	445 (59.4)	<0.001
Definitive vasculitis on temporal artery biopsy	335 (44.3)	1 (0.1)	<0.001
Halo sign on temporal artery ultrasound	211 (27.9)	1 (0.1)	<0.001
Bilateral axillary involvement on imaging§	57 (7.5)	12 (1.6)	<0.001
FDG-PET activity throughout aorta¶	52 (6.9)	9 (1.2)	<0.001

*Except where indicated otherwise, values are the number (%).
†Diagnoses of comparators for the classification criteria for giant cell arteritis (GCA) included Takayasu arteritis (n=251), Behçet's disease (n=133), polyarteritis nodosa (n=74), isolated aortitis (n=16), primary central nervous system vasculitis (n=16), large-vessel vasculitis (LVV) that could not be subtyped (n=9), other diseases that mimic LVV (n=250).
‡Absent or diminished pulse, tenderness or hard 'cord-like' appearance.
§Defined as damage (ie, stenosis, occlusion or aneurysm) on angiography (CT, MR or catheter based) or ultrasound, halo sign on ultrasound or abnormal FDG uptake on PET.
¶Descending thoracic and abdominal aorta.
CRP, C reactive protein; ESR, erythrocyte sedimentation rate; FDG-PET, ^{18}F-fluorodeoxyglucose–positron emission tomography; GCA, giant cell arteritis.

Validation of the final classification criteria for GCA

Using a cut-off of ≥6 in total risk score in the validation data set (see online supplemental appendix 18A for different cut-off points), the sensitivity was 87.0% (95% CI 82.0% to 91.0%) and specificity was 94.8% (95% CI 91.0% to 97.4%). The area under the curve for the model was 0.91 (95% CI 0.88 to 0.94) (online supplemental appendix 19A). The final 2022 ACR/EULAR classification criteria for GCA are presented in figure 1 (for the slide presentation versions, see online supplemental figure 1).

The performance characteristics of the criteria in different subsets of patients with GCA are shown in table 2 and online supplemental appendix 20A. Biopsy-proven GCA showed a sensitivity of 100% (95% CI 99.0% to 100.0%) and a specificity of 94.9% (95% CI 93.1% to 96.4%) and large-vessel GCA showed a sensitivity of 55.7% (95% CI 46.5% to 64.6%) and a specificity of 94.9% (95% CI 93.1% to 96.4%). Sensitivity of the

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CLASSIFICATION CRITERIA FOR **GIANT CELL ARTERITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- These classification criteria should be applied to classify the patient as having giant cell arteritis when a diagnosis of medium-vessel or large-vessel vasculitis has been made
- Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria

ABSOLUTE REQUIREMENT

Age ≥ 50 years at time of diagnosis	

ADDITIONAL CLINICAL CRITERIA

Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]	+3
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

Sum the scores for 10 items, if present. A score of ≥ 6 points is needed for the classification of GIANT CELL ARTERITIS.

1. Examination of the temporal artery showing absent or diminished pulse, tenderness, or hard 'cord-like' appearance.
2. Maximum erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) values prior to initiation of treatment for vasculitis.
3. Presence of either definitive vasculitis on temporal artery biopsy or halo sign on temporal artery ultrasound. There are no specific histopathologic criteria to define definitive vasculitis on temporal artery biopsy. Presence of giant cells, mononuclear leukocyte infiltration, and fragmentation of the internal elastic lamina were independently associated with histopathologic interpretation of definite vasculitis in the DCVAS cohort[24]. Halo sign is defined by the presence of an homogenous, hypoechoic wall thickening on ultrasound [25].
4. Bilateral axillary involvement is defined as luminal damage (stenosis, occlusion, or aneurysm) on angiography (computed tomography, magnetic resonance, or catheter-based) or ultrasound, halo sign on ultrasound, or fluorodeoxyglucose uptake on positron emission tomography.
5. Abnormal fluorodeoxyglucose (FDG) uptake in the arterial wall (e.g., greater than liver uptake by visual inspection) throughout the descending thoracic and abdominal aorta on positron emission tomography (PET).

Figure 1 The final 2022 American College of Rheumatology/EULAR Classification Criteria for Giant Cell Arteritis.

Table 2 Performance characteristics of the 2022 ACR/EULAR classification criteria for giant cell arteritis*

Patient subset	Total no patients (no GCA patients)	Sensitivity (95% CI)	Specificity (95% CI)	AUC (95% CI)
Development data set	1054 (518)	84.8 (81.4 to 87.7)	95.0 (92.8 to 96.7)	0.90 (0.88 to 0.92)
Validation data set	451 (238)	87.0 (82.0 to 91.0)	94.8 (91.0 to 97.4)	0.91 (0.88 to 0.94)
Biopsy-proven GCA†	1104 (355)	100.0 (99.0 to 100.0)	94.9 (93.1 to 96.4)	0.97 (0.97 to 0.98)
Large-vessel GCA‡	873 (124)	55.7 (46.5 to 64.6)	94.9 (93.1 to 96.4)	0.75 (0.71 to 0.80)

*Performance characteristics were tested in the subsets using the combined development and validation data sets to maximise sample size.
†Definite vasculitis on temporal artery biopsy (TAB).
‡Involvement of the aorta or its branch arteries on imaging, without vasculitis on TAB.
ACR, American College of Rheumatology; AUC, area under the curve; GCA, giant cell arteritis.

criteria in North America was 77.8% (95% CI 67.8% to 85.9%) and in Europe was 87.2% (95% CI 84.4% to 89.7%). Specificity in North America was 95.6% (95% CI 90.6% to 98.4%) and in Europe was 88.8% (95% CI 84.9% to 92.0%).

When the 1990 ACR classification criteria for GCA were applied to the DCVAS validation data set, the criteria performed poorly due to low sensitivity (80.3% (95% CI 74.6% to 85.1%)) but retained good specificity (92.5% (95% CI 88.1% to 95.7%)). In particular, the 1990 ACR criteria had poor sensitivity for patients with large-vessel GCA (37.1% (95% CI 28.6% to 46.2%)).

Age restrictions are absolute requirements for the 2022 ACR/EULAR classification criteria for GCA (≥50 years at diagnosis) and TAK (≤60 years at diagnosis). However, of the 70 patients with GCA diagnosed between the ages of 50 and 60 years, 44 (62.9%) met the new GCA classification criteria, 9 (12.9%) met the new TAK classification criteria, and only 2 (2.9%) met both the new GCA and TAK classification criteria (online supplemental appendix 21).

DISCUSSION

Presented here are the final 2022 ACR/EULAR GCA classification criteria. A six-stage approach was used, underpinned by data from the multinational, prospective DCVAS study and informed by expert review and consensus at each stage. The comparator group for developing and validating the criteria were other vasculitides and conditions that mimic GCA, where discrimination from GCA is difficult but important. In the validation set, the new criteria had a sensitivity of 87.0% (95% CI 82.0% to 91.0%) and a specificity of 94.8% (95% CI 91.0% to 97.4%). These are the official final values that should be quoted when referring to the criteria. The sensitivity and specificity values calculated in the development set were very similar, providing reassurance that the statistical methods avoided overfitting of models. The new criteria incorporate modern imaging techniques and have excellent specificity and sensitivity within a large, international cohort of patients with GCA. The criteria were designed to have face and content validity for use in clinical trials and other research studies.

These criteria are validated and intended for the purpose of *classification* of vasculitis and are not appropriate for use to establish a *diagnosis* of vasculitis. The aim of the classification criteria is to differentiate cases of GCA from similar types of vasculitis in research settings.[21] *Therefore, the criteria should only be applied when a diagnosis of LVV or medium-vessel vasculitis has been made and all potential "vasculitis mimics" have been excluded.* The exclusion of mimics is a key aspect of many classification criteria including those for Sjögren's syndrome[22] and rheumatoid arthritis.[16] The 1990 ACR classification criteria for vasculitis perform poorly when used for diagnosis (ie, when used to differentiate between cases of vasculitis vs mimics without vasculitis),[23] and it is expected that the 2022 criteria would also perform poorly if used inappropriately as diagnostic criteria in people for whom alternative diagnoses, such as infection or other non-vasculitis inflammatory diseases, are still being considered.

The 2022 ACR/EULAR GCA classification criteria are the result of an incredibly large worldwide effort, in which an extensive set of data was collected from >1000 patients with the submitted diagnosis of GCA. These criteria reflect current clinical practice, integrating different investigative methods (eg, TAB, ultrasound, angiography, PET) from various countries and medical specialties. Real cases of GCA and comparators were reviewed by a wide range of experts in vasculitis to establish an unbiased diagnostic reference to derive the criteria. Advanced statistical methods including lasso logistic regression and cluster analyses were applied, which facilitated testing for different covariates of interest, namely specific patterns of vasculitic involvement in imaging. Modern classification techniques with weighted criterion with threshold scores were used, allowing for more discriminatory items to factor more heavily in disease classification.

When compared with the original 1990 ACR classification criteria for GCA, the 2022 ACR/EULAR GCA classification criteria demonstrated greater sensitivity while maintaining similar specificity to the 1990 criteria. In particular, the new criteria were able to correctly classify more patients with the large-vessel GCA subtype, in whom the sensitivity of the 1990 ACR criteria was only 37.1%. Unlike the 1990 ACR criteria, an age of ≥50 years at diagnosis is a mandatory requirement to classify GCA in the 2022 ACR/EULAR criteria. This age threshold included>99% of patients with the reference diagnosis of GCA. The new criteria maintain good discriminative ability for patients diagnosed between the ages of 50 and 60 years, the interval where the absolute age requirements for the 2022 ACR/EULAR criteria for GCA and for TAK can overlap.

A potential limitation of these criteria was the non-standardised acquisition of clinical and imaging data among patients with LVV and comparators (eg, not all patients underwent vascular examination of the temporal arteries, PET was not available in many centres treating patients with LVV, and TAB and/or ultrasound was not performed in all patients with suspected GCA, etc). However, this reflects existing differences in clinical practice, and the 11 items included in the criteria allow for a feasible evaluation of patients in any clinical setting. These criteria also provide flexibility for classifying a patient, regardless of the diagnostic assessment strategy employed by physicians. Definite vasculitis on TAB was defined by the submitting physician and did not undergo central review; ~20% of cases did not have specific histopathologic findings but were reported as 'definitive vasculitis on TAB' alone. Most patients were recruited from Europe and North America, with fewer patients from Asia and Oceania. The performance characteristics of the criteria should be further tested in other populations that were underrepresented in the DCVAS cohort and may have different clinical presentations of GCA.

The 2022 ACR/EULAR classification criteria for GCA are the product of a rigorous methodologic process that utilised an extensive data set generated by the work of a remarkable international group of collaborators. These criteria have been endorsed by the ACR and EULAR and are now ready for use in clinical research.

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Correction notice This article has been corrected since it published Online First. An amendment has been made to figure one in the line: LABORATORY, IMAGING, AND BIOPSY CRITERIA, in the subrow labeled Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter2. This correction has not been made in print.

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Correction: *2022 American College of Rheumatology/EULAR classification criteria for giant cell arteritis*

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The correction was made to figure one in the line: LABORATORY, IMAGING, AND BIOPSY CRITERIA, in the subrow labelled Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]. This correction has not been made in print.

Figure 1

2022 AMERICAN COLLEGE OF RHEUMATOLOGY / EULAR

CLASSIFICATION CRITERIA FOR **GIANT CELL ARTERITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify the patient as having giant cell arteritis when a diagnosis of medium-vessel or large-vessel vasculitis has been made**
- **Alternate diagnoses mimicking vasculitis should be excluded prior to applying the criteria**

ABSOLUTE REQUIREMENT

Age ≥ 50 years at time of diagnosis	

ADDITIONAL CLINICAL CRITERIA

Morning stiffness in shoulders/neck	+2
Sudden visual loss	+3
Jaw or tongue claudication	+2
New temporal headache	+2
Scalp tenderness	+2
Abnormal examination of the temporal artery[1]	+2

LABORATORY, IMAGING, AND BIOPSY CRITERIA

Maximum ESR ≥ 50 mm/hour or maximum CRP ≥ 10 mg/liter[2]	+3
Positive temporal artery biopsy or halo sign on temporal artery ultrasound[3]	+5
Bilateral axillary involvement[4]	+2
FDG-PET activity throughout aorta[5]	+2

Sum the scores for 10 items, if present. A score of ≥ 6 points is needed for the classification of GIANT CELL ARTERITIS.

1. Examination of the temporal artery showing absent or diminished pulse, tenderness, or hard 'cord-like' appearance.
2. Maximum erythrocyte sedimentation rate (ESR) or C-reactive protein (CRP) values prior to initiation of treatment for vasculitis.
3. Presence of either definitive vasculitis on temporal artery biopsy or halo sign on temporal artery ultrasound. There are no specific histopathologic criteria to define definitive vasculitis on temporal artery biopsy. Presence of giant cells, mononuclear leukocyte infiltration, and fragmentation of the internal elastic lamina were independently associated with histopathologic interpretation of definite vasculitis in the DCVAS cohort[24]. Halo sign is defined by the presence of an homogenous, hypoechoic wall thickening on ultrasound [25].
4. Bilateral axillary involvement is defined as luminal damage (stenosis, occlusion, or aneurysm) on angiography (computed tomography, magnetic resonance, or catheter-based) or ultrasound, halo sign on ultrasound, or fluorodeoxyglucose uptake on positron emission tomography.
5. Abnormal fluorodeoxyglucose (FDG) uptake in the arterial wall (e.g., greater than liver uptake by visual inspection) throughout the descending thoracic and abdominal aorta on positron emission tomography (PET).

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