2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis

Peter C Grayson ,[1] Cristina Ponte ,[2,3] Ravi Suppiah,[4] Joanna C Robson,[5,6] Katherine Bates Gribbons,[1] Andrew Judge ,[7,8,9] Anthea Craven ,[7] Sara Khalid,[7] Andrew Hutchings ,[10] Debashish Danda ,[11] Raashid A Luqmani ,[7] Richard A Watts ,[7,12] Peter A Merkel ,[13] For the DCVAS Study Group

► Additional supplemental material is published online only. To view, please visit the journal online (http://dx.doi.org/10.1136/ard-2022-223482).

For numbered affiliations see end of article.

Correspondence to
Professor Peter A Merkel, Division of Rheumatology, Department of Medicine, University of Pennsylvania, Philadelphia, Pennsylvania 19104, USA;
pmerkel@upenn.edu

This article is published simultaneously in *Arthritis & Rheumatology*.

Received 13 October 2022
Accepted 13 October 2022
Published Online First
9 November 2022

To cite: Grayson PC, Ponte C, Suppiah R, *et al*. *Ann Rheum Dis* 2022;**81**:1654–1660.

ABSTRACT

Objective To develop and validate new classification criteria for Takayasu arteritis (TAK).

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

Results The development data set consisted of 316 cases of TAK and 323 comparators. The validation data set consisted of an additional 146 cases of TAK and 127 comparators. Age ≤60 years at diagnosis and imaging evidence of large-vessel vasculitis were absolute requirements to classify a patient as having TAK. The final criteria items and weights were as follows: female sex (+1), angina (+2), limb claudication (+2), arterial bruit (+2), reduced upper extremity pulse (+2), reduced pulse or tenderness of a carotid artery (+2), blood pressure difference between arms of ≥20 mm Hg (+1), number of affected arterial territories (+1 to +3), paired artery involvement (+1) and abdominal aorta plus renal or mesenteric involvement (+3). A patient could be classified as having TAK with a cumulative score of ≥5 points. When these criteria were tested in the validation data set, the model area under the curve was 0.97 (95% CI 0.94 to 0.99) with a sensitivity of 93.8% (95% CI 88.6% to 97.1%) and specificity of 99.2% (95% CI 96.7% to 100.0%).

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

INTRODUCTION

Takayasu arteritis (TAK) is one of the major forms of large-vessel vasculitis (LVV).[1] TAK is a chronic disease defined by granulomatous inflammation affecting the aorta and its primary branches. Complications from vascular damage can result in substantial morbidity including stroke, myocardial infarction, mesenteric ischaemia and limb claudication.

Unlike diagnostic criteria, the purpose of classification criteria is to ensure that a homogeneous population is selected for inclusion into clinical trials and other research studies.[2] In 1990, the American College of Rheumatology (ACR) endorsed classification criteria for TAK.[3] These criteria were developed using data from only 63 patients with TAK and have never been independently validated. In addition, these criteria were derived using data from patients exclusively from North America without representation from Europe or Asia, where clinical patterns of disease may differ,[4] limiting the generalisability of results. Given these constraints, the 1990 ACR criteria for TAK no longer satisfy accepted current standards[5] for classification criteria development, and updated criteria are warranted. Further highlighting a need for uniform, revised criteria in TAK is the use of divergent eligibility criteria to define study populations in two recent randomised clinical trials conducted in North America and Japan, making comparisons between the trial findings difficult.[6 7]

Advancements in imaging techniques and the ongoing adoption of noninvasive vascular imaging approaches in clinical practice have broadened understanding of the clinical heterogeneity in LVV.[8] Disease of the extracranial arteries is increasingly recognised in patients with giant cell arteritis (GCA), making the distinction between TAK and GCA more challenging.[9] Age is typically used as a primary classifier to differentiate between TAK and GCA; however, specific age thresholds to define each disease have not been standardised. Therefore, in addition to incorporating data from a larger patient population from a wider geographical spectrum, the updated TAK classification criteria should reflect modern clinical practice, including current imaging techniques, and also define specific age thresholds.

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

METHODS

An international Steering Committee comprising clinician investigators with expertise in vasculitis, statisticians and data managers was established to oversee the overall development of classification criteria for primary vasculitis.[10] A detailed and complete description of the methods involved in the development and validation of the classification criteria for TAK 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 (DCVAS) prospective observational study

A prospective, international, multisite observational study was conducted (see Appendix A 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 and atherosclerosis). Patients with TAK could only be enrolled within 5 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 geographical locations and specialties (see Appendix A) 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 LVV. Density plots were assessed to study age distribution at diagnosis and symptom onset for TAK and GCA. Absolute age requirements vs incorporation of age as a candidate criteria item were 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 were defined by K-means clustering.[11]

Stage 5: derivation of the final classification criteria for TAK

The DCVAS data set was split into development (70%) and validation (30%) sets. Comparisons were performed between cases of TAK and a randomly selected comparator group in the following proportions: GCA, 33.6%; other vasculitides that mimic GCA and TAK (isolated aortitis, primary central nervous system vasculitis, polyarteritis nodosa, Behçet's disease and other LVV), 33.1%; a comparator mimic of LVV (eg, headache syndrome or atherosclerosis), 33.3%. 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. 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 TAK

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 TAK using data from the combined development and validation sets, to maximise sample sizes for the subgroups. Patients were studied according to different intervals of age at diagnosis to determine if the criteria performed well across the age spectrum of TAK. Performance characteristics of the new criteria were also tested in patients recruited into the DCVAS study from different regions of the world where prevalence of TAK and clinical assessment approaches may differ. Comparison was made between the measurement properties of the new 2022 ACR/EULAR classification criteria for TAK and the 1990 ACR classification criteria.

RESULTS

Generation of candidate classification items for the systemic vasculitides

The Steering Committee identified >1000 candidate items for the DCVAS Case Report Form (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 TAK was confirmed in 500 of 610 cases (82.0%) after both expert panel reviews. The reasons for exclusion were diagnosis of TAK categorised as 'uncertain' or 'very uncertain' during panel review (n=95) or change in diagnosis during panel review to another type of vasculitis (eg, GCA, isolated aortitis, LVV that could not be subtyped) (n=10) or to a comparator disease (n=5). An additional 9 patients who were not initially diagnosed as having TAK by the submitting physician were diagnosed as having TAK after panel review and DCVAS Steering Committee member adjudication. Per Steering Committee consensus, imaging evidence of LVV was considered an absolute requirement to classify TAK. Of 509 cases confirmed by expert panel review, 47 patients with TAK did not have documented disease

according to a vascular imaging study and were excluded from further analysis, leaving a total of 462 patients with TAK for subsequent analysis.

Refinement of candidate items specifically for TAK.

Patients with TAK were diagnosed in the following age groups: 18–39 years (n=355; 77%); 40–60 years (n=104; 23%); and >60 years (n=3; <1%) (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 ≤60 years at diagnosis was considered an absolute requirement to classify a patient as having TAK.

Prevalence of arterial damage (stenosis, occlusion or aneurysm) was greater in TAK compared with GCA in the following nine arterial territories: thoracic aorta, abdominal aorta, left and right carotid, left and right subclavian, mesenteric and left and right renal arteries (online supplemental appendix 11). Therefore, a composite variable representing the number of affected arteries was created based on luminal damage in those nine territories. As previously reported, cluster analyses identified vascular damage in the abdominal aorta and the renal or mesenteric arteries as a specific imaging pattern for TAK in the DCVAS cohort[11]; thus, this arterial pattern was tested as a potential classifier of TAK (online supplemental appendix 12). Symmetric disease in branch arteries (carotid, subclavian and renal arteries) was seen in 30.3% patients with TAK compared with 2.7% of the comparators (p<0.01), and therefore, was included as a potential classifier. A systolic blood pressure difference of ≥20 mm Hg between upper extremities optimised specificity to differentiate TAK from other forms of LVV.

Following a data-driven and expert consensus process, 72 items from the DCVAS Case Report Form were retained for lasso regression analysis, including 32 demographic and clinical items, 14 laboratory items (including values of C reactive protein level and erythrocyte sedimentation rate, 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 arterial biopsy item (online supplemental appendix 13). Criteria for TAK and GCA were independently derived from this common set of 72 items.

Derivation of the final classification criteria for TAK.

Table 1 lists the demographic and disease features of the 462 patients with TAK and 450 comparators used to develop and validate the criteria, of which 316 patients with TAK and 323 comparators were in the development data set and 146 patients with TAK and 127 comparators were in the validation data set. The patients with TAK were recruited from Asia (n=298), Europe (n=130), North America (n=28), Africa (n=3) and Oceania (n=3). Clinical diagnoses assigned to patients in the comparator group are detailed in online supplemental appendix 14.

Lasso logistic regression analysis using all 72 items resulted in a model of 9 independent items (online supplemental appendix 15B). Weighting of individual criterion was based on logistic regression fitted to the nine selected predictors. The number of affected arterial territories functioned as an almost perfect classifier (online supplemental appendix 16B) and was thus also included in the final model, with criterion weighting determined by consensus of the Steering Committee (online supplemental appendix 17B).

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

	TAK (n=462)	Comparators (n=450)†	P value
Age, mean±SD years	32.3±10.4	58.6±18.0	<0.001
Female sex	391 (84.6)	246 (54.7)	<0.001
Clinical features			
Angina	56 (12.1)	7 (1.6)	<0.001
Arm claudication	233 (50.4)	11 (2.4)	<0.001
Leg claudication	88 (19.0)	17 (3.8)	<0.001
Vascular examination findings			
Arterial bruit	263 (56.9)	32 (7.1)	<0.001
Reduced or absent pulse in upper extremity	309 (66.9)	309 (66.9)	<0.001
Carotid artery with reduced pulse or tenderness	171 (37.0)	16 (3.6)	<0.001
Difference in systolic blood pressure ≥20 mm Hg between arms	190 (41.1)	16 (3.6)	<0.001
Imaging findings			
1 affected arterial territory	76 (16.5)	36 (8.0)	<0.001
2 affected arterial territories	114 (24.7)	12 (2.7)	<0.001
≥3 affected arterial territories	89 (19.2)	5 (1.1)	<0.001
Vasculitis affecting paired branch arteries	140 (30.3)	12 (2.7)	<0.001
Abdominal aorta involvement with renal or mesenteric artery involvement	83 (18.0)	5 (1.1)	<0.001

*Except where indicated otherwise, values are the number (%).
†Diagnoses of comparators for the classification criteria for TAK included giant cell arteritis (n=151), Behçet's disease (n=80), polyarteritis nodosa (n=39), clinically isolated aortitis (n=12), primary central nervous system vasculitis (n=11), large-vessel vasculitis (LVV) that could not be subtyped (n=7) and other diseases that mimic LVV (n=150).
TAK, Takayasu arteritis.

Validation of the final classification criteria for TAK

Using a cut-off of ≥5 in total risk score in the validation data set (see online supplemental appendix 18B for cut-off points), the sensitivity was 93.8% (95% CI 88.6% to 97.1%), and the specificity was 99.2% (95% CI 96.7% to 100.0%). The area under the curve for the model was 0.97 (95% CI 0.94 to 0.99) (online supplemental appendix 19B). The final classification criteria for TAK are shown 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 TAK are shown in table 2 and online supplemental appendix 20B. For patients who were diagnosed between 18 and 39 years of age, the sensitivity of the criteria was 94.0% (95% CI 91.0% to 96.3%), and the specificity was 97.7% (95% CI 91.9% to 99.7%). For patients who were diagnosed between 40 and 60 years of age, the sensitivity of the criteria was 83.7% (95% CI 75.1% to 90.2%) and the specificity was 91.8% (95% CI 85.4% to 96.0%). Because age restrictions are absolute requirements for the 2022 ACR/EULAR classification criteria for TAK (≤60 years at diagnosis) and GCA (≥50 years at diagnosis), patients with LVV between the ages of 50 and 60 years are potentially eligible to fulfil criteria for TAK and GCA. Of the 26 patients with TAK diagnosed between the ages of 50 and 60 years, 23 (88.5%) were classified correctly as having TAK, 1 (3.9%) was incorrectly classified as having GCA, and 1 (3.9%) fulfilled criteria for both TAK and GCA (online supplemental appendix 21). The criteria performed well in both

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CLASSIFICATION CRITERIA FOR **TAKAYASU ARTERITIS**

CONSIDERATIONS WHEN APPLYING THESE CRITERIA

- **These classification criteria should be applied to classify the patient as having Takayasu 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 REQUIREMENTS

Age ≤ 60 years at time of diagnosis	
Evidence of vasculitis on imaging[1]	

ADDITIONAL CLINICAL CRITERIA

Female sex	+1
Angina or ischemic cardiac pain	+2
Arm or leg claudication	+2
Vascular bruit[2]	+2
Reduced pulse in upper extremity[3]	+2
Carotid artery abnormality[4]	+2
Systolic blood pressure difference in arms ≥ 20 mm Hg	+1

ADDITIONAL IMAGING CRITERIA

Number of affected arterial territories (select one)[5]	
One arterial territory	+1
Two arterial territories	+2
Three or more arterial territories	+3
Symmetric involvement of paired arteries[6]	+1
Abdominal aorta involvement with renal or mesenteric involvement[7]	+3

Sum the scores for 10 items, if present. A score of ≥ 5 points is needed for the classification of TAKAYASU ARTERITIS.

1. Evidence of vasculitis in the aorta or branch arteries must be confirmed by vascular imaging (e.g., computed tomographic/catheter-based/magnetic resonance angiography, ultrasound, positron emission tomography).
2. Bruit detected by auscultation of a large artery, including the aorta, carotid, subclavian, axillary, brachial, renal, or iliofemoral arteries.
3. Reduction or absence of pulse by physical examination of the axillary, brachial, or radial arteries.
4. Reduction or absence of pulse of the carotid artery or tenderness of the carotid artery.
5. Number of arterial territories with luminal damage (e.g., stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography from the following nine territories: thoracic aorta, abdominal aorta, mesenteric, left or right carotid, left or right subclavian, left or right renal arteries.
6. Bilateral luminal damage (stenosis, occlusion, or aneurysm) detected by angiography or ultrasonography in any of the following paired vascular territories: carotid, subclavian, or renal arteries.
7. Luminal damage (stenosis, occlusion, aneurysm) detected by angiography or ultrasonography involving the abdominal aorta and either the renal or mesenteric arteries.

Figure 1 The final 2022 American College of Rheumatology/EULAR classification criteria for Takayasu arteritis.

Asia (sensitivity 92.0%, specificity 93.2%) and Europe/North America (sensitivity 90.5%, specificity 94.4%).

When the 1990 ACR classification criteria for TAK were applied to the DCVAS validation data set, the criteria performed poorly due to low sensitivity (84.3% (95% CI 77.3% to 89.7%)) but retained excellent specificity (99.2% (95% CI 95.7% to 100.0%)). In particular, the 1990 criteria had poor sensitivity for patients who were diagnosed as having TAK between 40 and 60 years of age (62.5% (95% CI 52.5% to 71.8%)).

DISCUSSION

Presented here are the final 2022 ACR/EULAR TAK 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 TAK, where discrimination from TAK is difficult but important. In the validation

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

Patient subset	Total no patients (no TAK patients)	Sensitivity (95% CI)	Specificity (95% CI)	AUC (95% CI)
Development data set	639 (316)	89.9 (86.0 to 93.0)	96.6 (94.0 to 98.3)	0.93 (0.91 to 0.95)
Validation data set	273 (146)	93.8 (88.6 to 97.1)	99.2 (96.7 to 100.0)	0.97 (0.94 to 0.99)
Age intervals				
18–39 years	437 (351)	94.0 (91.0 to 96.3)	97.7 (91.9 to 99.7)	0.96 (0.94 to 0.98)
40–60 years	226 (104)	83.7 (75.1 to 90.2)	91.8 (85.4 to 96.0)	0.88 (0.83 to 0.92)
World regions				
North America	127 (28)	85.7 (67.3 to 96.0)	92.9 (86.0 to 97.1)	0.89 (0.82 to 0.96)
Europe	422 (130)	91.5 (85.4 to 95.7)	94.9 (91.7 to 97.1)	0.93 (0.90 to 0.96)
North America/Europe combined	549 (158)	90.5 (84.8 to 94.6)	94.4 (91.6 to 96.4)	0.92 (0.90 to 0.95)
Asia	357 (298)	92.0 (88.3 to 94.8)	93.2 (83.5 to 98.1)	0.94 (0.89 to 0.96)

*Performance characteristics for the age and regional subsets were reported using data from the combined development and validation data sets to maximise sample size.
ACR, American College of Rheumatology; AUC, area under the curve; TAK, Takayasu arteritis.

data set, the new criteria had a sensitivity of 93.8% (95% CI 88.6% to 97.1%) and a specificity of 99.2% (95% CI 96.7% to 100.0%). These are the official final values that should be quoted when referring to the criteria. The sensitivity and specificity values calculated in the development data set were very similar, providing reassurance that the statistical methods avoided overfitting of models. Calculations of sensitivity and specificity for patient subgroups were made in the combined development and validation data sets to maximise sample sizes for the subgroups. Reassuringly, the new criteria for TAK have excellent sensitivity and specificity across different regions of the world. The criteria also incorporate modern imaging techniques, which are useful both to diagnose LVV and to differentiate among different types of vasculitis. 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.[2] The aim of the classification criteria is to differentiate cases of TAK from similar types of vasculitis in research settings.[5] *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.* For example, the criteria were not developed to differentiate patients with TAK from patients with atherosclerosis or noninflammatory genetic diseases that damage the large arteries. 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), and it is expected that the 2022 criteria would also perform poorly if used inappropriately as diagnostic criteria.[12]

The 2022 ACR/EULAR TAK classification criteria reflect the collaborative effort of the international vasculitis community to delineate the salient clinical features that differentiate TAK from other forms of vasculitis, most notably GCA. The final criteria include 10 clinical items that are routinely assessed during clinical evaluation of patients with TAK. The criteria highlight the importance of clinical symptoms, vascular physical examination and vascular imaging as important disease classifiers. Features of TAK may differ in patients from different parts of the world.[13] The 2022 ACR/EULAR TAK classification criteria retained excellent performance characteristics when tested in patients from different regions, including Asia where the disease is most prevalent.[14] While TAK is often considered a disease of the young, 25% of the patients with TAK in the DCVAS cohort were older than 40 years at the time of diagnosis. Therefore, an age at diagnosis of ≤60 years, rather than a lower age threshold, was set as an absolute requirement for disease classification. The 2022 ACR/EULAR TAK classification criteria performed well when applied to patients ages 18–60 years and outperformed the 1990 ACR Classification Criteria for TAK in the subset of patients diagnosed as having TAK ages 40–60 years.

There are several strengths of the new 2022 ACR/EULAR TAK classification criteria. The criteria were developed by a large group of international experts in systemic vasculitis, with guidance from the ACR regarding modern methods of classification criteria development. The criteria represent several important methodologic advancements compared with the original 1990 ACR classification criteria for TAK. First, expert review rather than submitting physician diagnosis was used as the diagnostic reference standard to minimise investigator bias. Second, while the 1990 ACR criteria were entirely derived using data from 63 North American patients with TAK and not validated in a separate data set, the new criteria were developed in 316 patients with TAK and validated in an independent data set which contained an additional 146 patients with TAK from an international cohort. Third, unlike the 1990 ACR criteria, the new ACR/EULAR TAK criteria are weighted to reflect the relative importance of specific items (eg, number of affected arterial territories). Finally, when both criteria sets were tested within the DCVAS cohort, the performance characteristics of the 2022 ACR/EULAR TAK criteria outperformed the 1990 ACR criteria.

There are some study limitations to consider. Acquisition of clinical and imaging data among patients with LVV and comparators was not standardised (eg, not all pulses were recorded by the investigators; patients with suspected diagnosis of TAK rarely underwent investigation of the cranial arteries; temporal artery biopsy was not performed in all patients with suspected GCA). However, this limitation reflects the existing differences in how these diseases are assessed in routine clinical practice. Most patients were recruited from Europe, Asia and North America, with fewer patients from Africa and Oceania. The performance characteristics of the criteria should be further tested in populations that were underrepresented in the DCVAS cohort and may have different clinical presentations of TAK. These criteria were developed using data collected from adult patients with vasculitis and should be tested in children with TAK.[15]

The 2022 ACR/EULAR classification criteria for TAK are the product of a rigorous methodologic process that used 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 research.

Author affiliations
[1]Systemic Autoimmunity Branch, National Institute of Arthritis and Musculoskeletal and Skin Diseases, NIH, Bethesda, Maryland, USA
[2]Department of Rheumatology, Centro Hospitalar Universitário Lisboa Norte, Centro Académico de Medicina de Lisboa, Lisbon, Portugal
[3]Rheumatology Research Unit, Instituto de Medicina Molecular, Faculdade de Medicina, Universidade de Lisboa, Centro Académico de Medicina de Lisboa, Lisbon, Portugal
[4]Te Whatu Ora - Health New Zealand, Auckland, New Zealand
[5]Centre for Health and Clinical Research, University of the West of England, Bristol, UK
[6]Rheumatology Department, University Hospitals Bristol and Weston NHS Foundation Trust, Bristol, UK
[7]Nuffield Department of Orthopaedics Rheumatology and Musculoskeletal Sciences, Oxford NIHR Biomedical Research Centre, University of Oxford, Oxford, UK
[8]Musculoskeletal Research Unit, Translational Health Sciences, Bristol Medical School, University of Bristol, Bristol, UK
[9]National Institute for Health Research Bristol Biomedical Research Centre, University Hospitals Bristol and Weston NHS Foundation Trust and University of Bristol, Bristol, UK
[10]Department of Health Services Research and Policy, London School of Hygiene and Tropical Medicine, London, UK
[11]Department of Clinical Immunology and Rheumatology, Christian Medical College, Vellore, Tamil Nadu, India
[12]Norwich Medical School, University of East Anglia, Norwich, UK
[13]Division of Rheumatology, Department of Medicine, and Division of Epidemiology, Department of Biostatistics, Epidemiology, and Informatics, University of Pennsylvania, Philadelphia, Pennsylvania, USA

Acknowledgements We acknowledge the patients and clinicians who provided data to the DCVAS project.

Collaborators The DCVAS study investigators are as follows: Paul Gatenby (ANU Medical Centre, Canberra, Australia); Catherine Hill (Central Adelaide Local Health Network: The Queen Elizabeth Hospital, Australia); Dwarakanathan Ranganathan (Royal Brisbane and Women's Hospital, Australia); Andreas Kronbichler (Medical University Innsbruck, Austria); Daniel Blockmans (University Hospitals Leuven, Belgium); Lillian Barra (Lawson Health Research Institute, London, Ontario, Canada); Simon Carette, Christian Pagnoux (Mount Sinai Hospital, Toronto, Canada); Navjot Dhindsa (University of Manitoba, Winnipeg, Canada); Aurore Fifi-Mah (University of Calgary, Alberta, Canada); Nader Khalidi (St Joseph's Healthcare, Hamilton, Ontario, Canada); Patrick Liang (Sherbrooke University Hospital Centre, Canada); Nataliya Milman (University of Ottawa, Canada); Christian Pineau (McGill University, Canada); Xinping Tian (Peking Union Medical College Hospital, Beijing, China); Guochun Wang (China-Japan Friendship Hospital, Beijing, China); Tian Wang (Anzhen Hospital, Capital Medical University, China); Ming-hui Zhao (Peking University First Hospital, China); Vladimir Tesar (General University Hospital, Prague, Czech Republic); Bo Baslund (University Hospital, Copenhagen [Rigshospitalet], Denmark); Nevin Hammam (Assiut University, Egypt); Amira Shahin (Cairo University, Egypt); Laura Pirila (Turku University Hospital, Finland); Jukka Putaala (Helsinki University Central Hospital, Finland); Bernhard Hellmich (Kreiskliniken Esslingen, Germany); Jörg Henes (Universitätsklinikum Tübingen, Germany); Julia Holle, Frank Moosig (Klinikum Bad Bramstedt, Germany); Peter Lamprecht (University of Lübeck, Germany); Thomas Neumann (Universitätsklinikum Jena, Germany); Wolfgang Schmidt (Immanuel Krankenhaus Berlin, Germany); Cord Sunderkoetter (Universitätsklinikum Müenster, Germany); Zoltan Szekanecz (University of Debrecen Medical and Health Science Center, Hungary); Debashish Danda (Christian Medical College & Hospital, Vellore, India); Siddharth Das (Chatrapathi Shahuji Maharaj Medical Center, Lucknow [IP], India); Rajiva Gupta (Medanta, Delhi, India); Liza Rajasekhar (NIMS, Hyderabad, India); Aman Sharma (Postgraduate Institute of Medical Education and Research, Chandigarh, India); Shrikant Wagh (Jehangir Clinical Development Centre, Pune [IP], India); Michael Clarkson (Cork University Hospital, Ireland); Eamonn Molloy (St. Vincent's University Hospital, Dublin, Ireland); Carlo Salvarani (Santa Maria Nuova Hospital, Reggio Emilia, Italy); Franco Schiavon (L'Azienda Ospedaliera of University of Padua, Italy); Enrico Tombetti (Università Vita- Salute San Raffaele Milano, Italy); Augusto Vaglio (University of Parma, Italy); Koichi Amano (Saitama Medical University, Japan); Yoshihiro Arimura (Kyorin University Hospital, Japan); Hiroaki Dobashi (Kagawa University Hospital, Japan); Shouichi Fujimoto (Miyazaki University Hospital [HUB], Japan); Masayoshi Harigai, Fumio Hirano (Tokyo Medical and Dental University Hospital, Japan); Junichi Hirahashi (University Tokyo Hospital, Japan); Sakae Honma (Toho University Hospital, Japan); Tamihiro Kawakami (St. Marianna University Hospital Dermatology, Japan); Shigeto Kobayashi (Juntendo University Koshigaya Hospital, Japan); Hajime Kono (Teikyo University, Japan); Hirofumi Makino (Okayama University Hospital, Japan); Kazuo Matsui (Kameda Medical Centre, Kamogawa, Japan); Eri Muso (Kitano Hospital, Japan); Kazuo Suzuki, Kei Ikeda (Chiba University Hospital, Japan); Tsutomu Takeuchi (Keio University Hospital, Japan); Tatsuo Tsukamoto (Kyoto University Hospital, Japan); Shunya Uchida (Teikyo University Hospital, Japan); Takashi Wada (Kanazawa University Hospital, Japan); Hidehiro Yamada (St. Marianna University Hospital Internal Medicine, Japan); Kunihiro Yamagata (Tsukuba University Hospital, Japan); Wako Yumura (IUHW Hospital [Jichi Medical University Hospital], Japan); Kan Sow Lai (Penang General Hospital, Malaysia); Luis Felipe Flores-Suarez (Instituto Nacional de Enfermedades Respiratorias, Mexico City, Mexico); Andrea Hinojosa-Azaola (Instituto Nacional de Ciencias Médicas y Nutricion Salvador Zubiran, Mexico City, Mexico); Bram Rutgers (University Hospital Groningen, Netherlands); Paul-Peter Tak (Academic Medical Centre, University of Amsterdam, Netherlands); Rebecca Grainger (Wellington, Otago, New Zealand); Vicki Quincey (Waikato District Health Board, New Zealand); Lisa Stamp (University of Otago, Christchurch, New Zealand); Ravi Suppiah (Auckland District Health Board, New Zealand); Emilio Besada (Tromsø, Northern Norway, Norway); Andreas Diamantopoulos (Hospital of Southern Norway, Kristiansand, Norway); Jan Sznajd (University of Jagiellonian, Poland); Elsa Azevedo (Centro Hospitalar de Sao Joao, Porto, Portugal); Ruth Geraldes (Hospital de Santa Maria, Lisbon, Portugal); Miguel Rodrigues (Hospital Garcia de Orta, Almada, Portugal); Ernestina Santos (Hospital Santo Antonio, Porto, Portugal); Yeong-Wook Song (Seoul National University Hospital, Republic of Korea); Sergey Moiseev (First Moscow State Medical University, Russia); Alojzija Hocevar (University Medical Centre Ljubljana, Slovenia); Maria Cinta Cid (Hospital Clinic de Barcelona, Spain); Xavier Solanich Moreno (Hospital de Bellvitge-Idibell, Spain); Inoshi Atukorala (University of Colombo, Sri Lanka); Ewa Berglin (Umeå University Hospital, Sweden); Aladdin Mohammed (Lund-Malmo University, Sweden); Mårten Segelmark (Linköping University, Sweden); Thomas Daikeler (University Hospital Basel, Switzerland); Haner Direskeneli (Marmara University Medical School, Turkey); Gulen Hatemi (Istanbul University, Cerrahpasa Medical School, Turkey); Sevil Kamali (Istanbul University, Istanbul Medical School, Turkey); Ömer Karadag (Hacettepe University, Turkey); Seval Pehlevan (Fatih University Medical Faculty, Turkey); Matthew Adler (Frimley Health NHS Foundation Trust, Wexham Park Hospital, UK); Neil Basu (NHS Grampian, Aberdeen Royal Infirmary, UK); Iain Bruce (Manchester University Hospitals NHS Foundation Trust, UK); Kuntal Chakravarty (Barking, Havering and Redbridge University Hospitals NHS Trust, UK); Bhaskar Dasgupta (Southend University Hospital NHS Foundation Trust, UK); Oliver Flossmann (Royal Berkshire NHS Foundation Trust, UK); Nagui Gendi (Basildon and Thurrock University Hospitals NHS Foundation Trust, UK); Alaa Hassan (North Cumbria University Hospitals, UK); Rachel Hoyles (Oxford University Hospitals NHS Foundation Trust, UK); David Jayne (Cambridge University Hospitals NHS Foundation Trust, UK); Colin Jones (York Teaching Hospitals NHS Foundation Trust, UK); Rainer Klocke (The Dudley Group NHS Foundation Trust, UK); Peter Lanyon (Nottingham University Hospitals NHS Trust, UK); Cathy Laversuch (Taunton & Somerset NHS Foundation Trust, Musgrove Park Hospital, UK); Raashid Luqmani, Joanna Robson (Nuffield Orthopaedic Centre, Oxford, UK); Malgorzata Magliano (Buckinghamshire Healthcare NHS Trust, UK); Justin Mason (Imperial College Healthcare NHS Trust, UK); Win Win Maw (Mid Essex Hospital Services NHS Trust, UK); Iain McInnes (NHS Greater Glasgow & Clyde, Gartnavel Hospital & GRI, UK); John Mclaren (NHS Fife, Whyteman's Brae Hospital, UK); Matthew Morgan (University Hospitals Birmingham NHS Foundation Trust, Queen Elizabeth Hospital, UK); Ann Morgan (Leeds Teaching Hospitals NHS Trust, UK); Chetan Mukhtyar (Norfolk and Norwich University Hospitals NHS Foundation Trust, UK); Edmond O'Riordan (Salford Royal NHS Foundation Trust, UK); Sanjeev Patel (Epsom and St Helier University Hospitals NHS Trust, UK); Adrian Peall (Wye Valley NHS Trust, Hereford County Hospital, UK); Joanna Robson (University Hospitals Bristol NHS Foundation Trust, UK); Srinivasan Venkatachalam (The Royal Wolverhampton NHS Trust, UK); Erin Vermaak, Ajit Menon (Staffordshire & Stoke on Trent Partnership NHS Trust, Haywood Hospital, UK); Richard Watts (East Suffolk and North Essex NHS Foundation Trust, UK); Chee-Seng Yee (Doncaster and Bassetlaw Hospitals NHS Foundation Trust, UK); Daniel Albert (DartmouthHitchcock Medical Center, US); Leonard Calabrese (Cleveland Clinic Foundation, US); Sharon Chung (University of California, San Francisco, US); Lindsy Forbess (Cedars-Sinai Medical Center, US); Angelo Gaffo (University of Alabama at Birmingham, US); Ora Gewurz-Singer (University of Michigan, US); Peter Grayson (Boston University School of Medicine, US); Kimberly Liang (University of Pittsburgh, US); Eric Matteson (Mayo Clinic, US); Peter A. Merkel, Rennie Rhee (University of Pennsylvania, US); Jason Springer (University of Kansas Medical Center Research Institute, US); and Antoine Sreih (Rush University Medical Center, US).

Contributors All authors were involved in drafting the article or revising it critically for important intellectual content, and all authors approved the final versionto be published. PAM had full access to all of the data in the study and takes responsibility for the integrity of the data and the accuracy of the data analysis. Study conception and design. PCG, CP, RS, JCR, AJ, AC, AH, RAL, RAW and PAM. Acquisition of data. PCG, CP, RS, JCR, AC, DD, RAL, RAW and PAM. Analysis and interpretation of data. PCG, CP, RS, JCR, KBG, AJ, AC, SK, AH, RAL, RAW and PAM.

Funding The Diagnostic and Classification Criteria in Vasculitis (DCVAS) study, which included the development of this classification criteria, was funded by grants from the American College of Rheumatology (ACR), EULAR, the Vasculitis Foundation and the University of Pennsylvania Vasculitis Center. This study was also supported by the Intramural Research Program of the National Institute of Arthritis and Musculoskeletal and Skin Diseases, NIH.

Competing interests None declared.

Patient consent for publication Not applicable.

Ethics approval Ethical approval was obtained from local ethics committees.

Provenance and peer review Commissioned; internally peer reviewed.

ORCID iDs
Peter C Grayson http://orcid.org/0000-0002-8269-9438
Cristina Ponte http://orcid.org/0000-0002-3989-1192
Andrew Judge http://orcid.org/0000-0003-3015-0432
Anthea Craven http://orcid.org/0000-0001-9477-7889
Andrew Hutchings http://orcid.org/0000-0003-0215-9923
Debashish Danda http://orcid.org/0000-0002-2121-0942
Raashid A Luqmani http://orcid.org/0000-0002-4446-5841
Richard A Watts http://orcid.org/0000-0002-2846-4769
Peter A Merkel http://orcid.org/0000-0001-9284-7345

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