

## ORIGINAL RESEARCH

Performance in adults of the EULAR/  
PRINTO/PRES (Ankara 2008)  
classification criteria for IgA vasculitis

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**ABSTRACT**

**Objective** To examine the performance in adults of the European Alliance of Associations for Rheumatology (EULAR)/Pediatric Rheumatology European Society (PReS)-endorsed Ankara 2008 classification criteria for IgA vasculitis (IgAV).

**Methods** The EULAR/PReS/Ankara 2008 classification criteria for IgAV were applied to patients enrolled in an international observational cohort which included patients with IgAV and comparators with other forms of small-vessel and medium-vessel vasculitis. After the initial assessment of the performance of the criteria, possible revisions to increase the performance were tested. The revised criteria were then assessed in an independent validation cohort within a multicentre Turkish vasculitis registry.

**Results** The dataset consisted of 178 IgAV cases and 1705 comparators. The Ankara 2008 criteria require skin involvement plus one of the following four criteria: abdominal pain, a biopsy showing IgA deposition, arthritis or arthralgia, or renal involvement (any haematuria and/or proteinuria). The specificity of the criteria improved when a positive test for anti-neutrophil cytoplasmic autoantibody or blood cryoglobulins was considered an exclusion criterion. The revised criteria had a sensitivity of 76.4% (95% CI 69.8% to 82.2%) and a specificity of 94.5% (95.0% CI 93.4% to 95.1%). In the validation set, the sensitivity and specificity of the revised criteria were 97.8% (95% CI 94.0% to 99.0%) and 85.0% (95.0% CI 78.0% to 90.0%), respectively.

**Conclusion** The revised EULAR/PReS-endorsed Ankara 2008 IgAV classification criteria perform well in adults with IgAV and are appropriate for use in clinical research.

**INTRODUCTION**

IgA vasculitis (IgAV), formerly known as Henoch-Schönlein purpura (HSP), is the most common systemic vasculitis in childhood. IgAV is characterised by IgA1-dominant immune deposits affecting small vessels, predominantly capillaries, venules or

**WHAT IS ALREADY KNOWN ON THIS TOPIC**

⇒ The European Alliance of Associations for Rheumatology/Paediatric Rheumatology International Trials Organisation/Pediatric Rheumatology European Society Ankara 2008 classification criteria for IgA vasculitis (IgAV) were developed and validated only in paediatric populations, and no validated criteria exist for adult patients. This paper presents the results of work to validate the criteria in adults.

**WHAT THIS STUDY ADDS**

⇒ Adding positive tests for anti-neutrophil cytoplasmic autoantibody or blood cryoglobulins as exclusion criteria to the original Ankara 2008 criteria significantly improves specificity for IgAV in adults from 73.4% to 94.5%.

**HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY**

⇒ These new validated classification criteria for adult IgAV can be used in clinical research and enable standardising classification across the lifespan.

arterioles.<sup>1 2</sup> IgAV is less common in adults, with an estimated incidence of 1.3 per 100 000, compared with an annual incidence of 29.9 per 100 000 in children.<sup>2</sup> IgAV primarily affects the skin, gastrointestinal tract and joints but can also result in glomerulonephritis.<sup>1</sup>

The 1990 American College of Rheumatology (ACR) classification criteria for HSP (IgAV) were derived using data from 85 adult patients with IgAV and were never validated. These classification criteria required the presence of at least two of the following: (1) age ≤20 years at disease onset, (2) palpable purpura, (3) acute abdominal pain and (4) a biopsy showing granulocytes in the walls of

small arterioles/venules. However, these criteria rely on non-specific features and do not address the importance of IgA in the pathogenesis of disease.<sup>3</sup> Furthermore, the criteria were derived from a cohort in which 64% of patients were children.

In 2008, paediatricians developed and validated the European Alliance of Associations for Rheumatology (EULAR) and Pediatric Rheumatology European Society (PReS)-endorsed classification criteria for IgAV, also known as the Ankara 2008 criteria.<sup>4</sup> The Ankara 2008 criteria require the presence of purpura (mandatory) plus one of the following four criteria: (1) abdominal pain, (2) a biopsy showing IgA deposition, (3) arthritis or arthralgia and (4) renal involvement (any haematuria and/or proteinuria). The final analysis included 827 patients with IgAV, compared with 356 patients with other forms of vasculitis, resulting in a high sensitivity (100%) and specificity (87%) in children.

A study revisiting the performance of the ACR 1990 criteria in a large cohort of patients with vasculitis highlighted the need for updated classification criteria.<sup>5</sup> Neither the 1990 ACR criteria nor the paediatric Ankara 2008 criteria were validated in adults. Adult IgAV differs from childhood IgAV, with more systemic involvement (eg, more renal involvement) in adults. Many forms of small-vessel or medium-vessel vasculitis overlap clinically with IgAV and are more common in adults (eg, anti-neutrophil cytoplasmic autoantibody (ANCA)-associated vasculitis) than in children. Thus, there is a substantial need for criteria to differentiate IgAV from other vasculitides in adults.

The aim of the current study was to examine the performance of the Ankara 2008 criteria in adults with IgAV.

## METHODS

### Patients and inclusion criteria

This analysis used data from the Diagnosis and Classification of Vasculitis Study (DCVAS), a large international research initiative focused on revising the classification systems for different types of vasculitis. Such criteria for several forms of vasculitis have previously been reported from this cohort.<sup>6–11</sup> For the current study, data from DCVAS were included from (1) patients with a diagnosis of IgAV and (2) a set of comparators with polyarteritis nodosa (PAN), ANCA-associated vasculitis (AAV), cryoglobulinaemic vasculitis or a different small-vessel vasculitis, including clinical, serological and pathological data. The DCVAS project included patients evaluated by rheumatologists, nephrologists, dermatologists and internal medicine physicians. Diagnoses made by the submitting physicians were considered the gold standard. Submitting physicians stated their level of certainty regarding the diagnosis at baseline as follows: very certain (>75%), moderately certain (50–74%), uncertain (25–49%) and very uncertain (<25%). Only patients for whom the investigator was very certain (>75%) about the diagnosis were included in the analysis for both IgAV and control groups.

Patients for whom the investigator changed their level of diagnostic certainty at 6 months to <75% or had a change of diagnosis were excluded from the analysis for both groups. Patients with a diagnosis of IgAV with nephropathy but no extrarenal involvement were excluded from the analysis. Patients with a skin biopsy showing IgA deposition without clinical documentation of skin involvement were also excluded. This study was approved by the Institutional Review Boards/Ethics Committees at all participating sites, and written informed consent was obtained from all patients prior to enrolment. The study was conducted in accordance with good clinical practice and the principles of the Declaration of Helsinki.

### Analysis

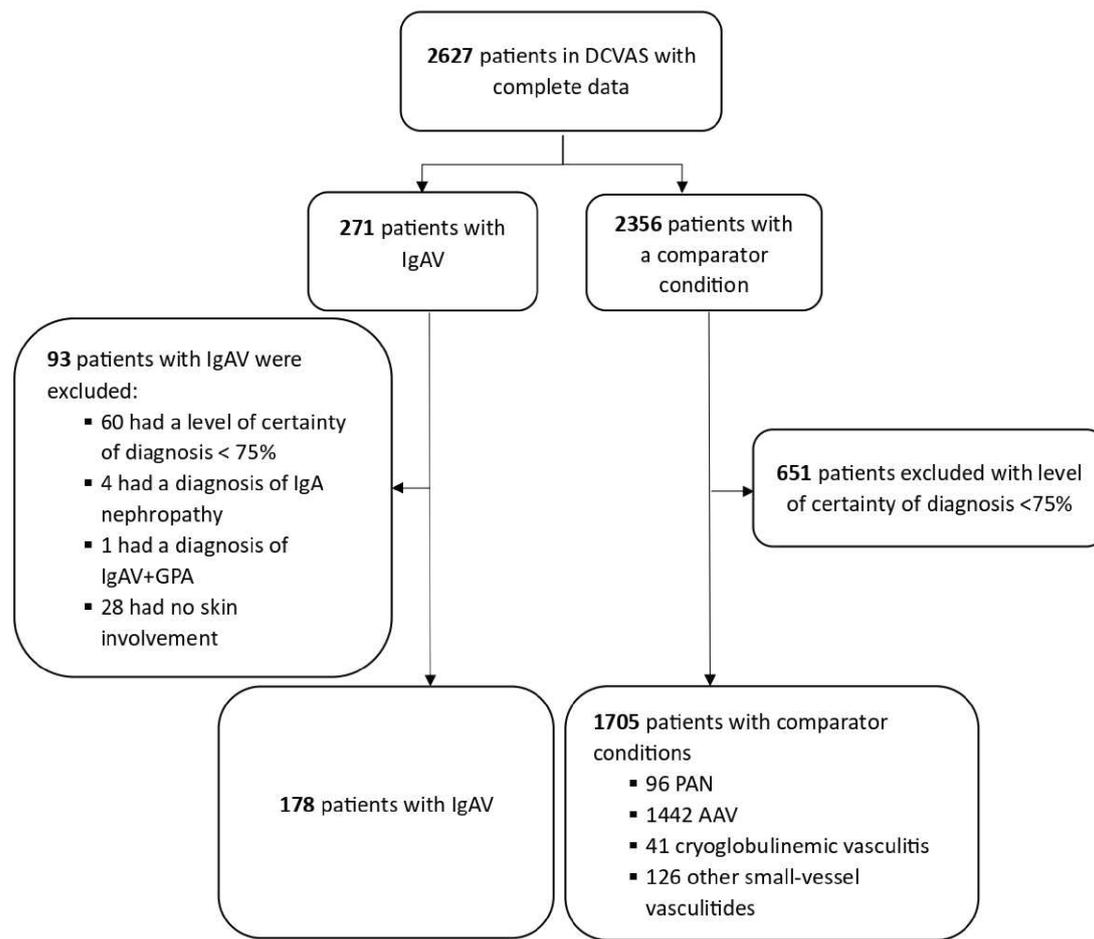
The performance of the original EULAR/PReS/Ankara 2008 criteria was analysed in the DCVAS cohort. Skin involvement plus one of the following four criteria was required: abdominal pain, a biopsy showing IgA deposition, arthritis or arthralgia, or renal involvement (any haematuria and/or proteinuria). Missing information was considered absent. The presence of each individual revised Ankara 2008 criterion was evaluated in each case in both the IgAV and comparator groups. With the goal of maximising specificity, revisions were considered, tested and subsequently validated in an independent cohort. The performance was also re-analysed after the Ankara 2008 criteria items were revised by adding positivity for serum ANCA and/or cryoglobulins as an exclusion criterion. The revised Ankara 2008 criteria were considered 'met' if the aforementioned criteria were fulfilled. The definitions for each criterion are shown in online supplemental table 1.

### Validation cohort

A revised version of the Ankara 2008 criteria that contained additional exclusion criteria was tested in an independent cohort of adult patients with IgAV from the Turkish Vasculitis Study Group (TRVaS) registry. TRVaS is a prospective, web-based registry enrolling all types of primary vasculitis between 2020 and 2024.<sup>12</sup> Diagnoses made by the submitting physicians were used as the gold standard. All patients included in the validation set had over 75% level of certainty for diagnosis.

### Statistical analysis

Statistical analyses were performed using the SPSS software V.25. Variables were investigated using visual (histogram, probability plots) and analytic methods (Kolmogorov-Smirnov) to determine whether they were normally distributed. Descriptive analyses were presented with proportions, medians, and minimum and maximum values, as appropriate. For comparisons of proportions between groups,  $\chi^2$  or Fisher's exact test was used, as appropriate. The Mann-Whitney U test was used to compare the non-normally distributed numeric variables between the two groups. The Wilcoxon test was used to analyse the differences between paired samples.



**Figure 1** Study population. AAV, anti-neutrophil cytoplasmic autoantibody-associated vasculitis; DCVAS, Diagnosis and Classification in Vasculitis Study; GPA, granulomatosis with polyangiitis; IgAV, IgA vasculitis; PAN, polyarteritis nodosa.

A p value of  $<0.05$  was considered statistically significant, and the CI was 95%.

The Ankara 2008 criteria and revised Ankara 2008 criteria were applied to the IgAV and comparator groups. For each criterion or classification definition, sensitivity (correct identification of patients with the specific disease, calculated as the number of true positives divided by the total number of patients with the disease) and specificity (correct identification of patients without the disease, calculated as the number of true negatives divided by the total number of patients without the disease) were determined. Receiver operating characteristic (ROC) curve analysis was conducted with the area under the ROC curve which compared the performance of these criteria in distinguishing between patients with IgAV and those with other forms of vasculitis.

## RESULTS

Data from 2627 patients were available, including 271 with IgAV and 2356 with comparator conditions. 744 patients (93 with IgAV; 651 with comparator conditions) were excluded from the analysis (figure 1). Thus, 178 patients with a physician-submitted diagnosis of IgAV were included in the analysis, with 1705 patients with other types of vasculitis included as comparators. The

comparator group was comprised of 96 patients (5.6%) with PAN, 1442 patients (84.5%) with AAV, 41 patients (2.4%) with cryoglobulinaemic vasculitis and 126 patients (7.4%) with other small-vessel vasculitides.

## Demographic and clinical characteristics

The demographic and clinical data for both groups are presented in table 1. The median age at onset was 51.5 years ( $\pm 21.1$ ) for the IgAV group and 54.1 years ( $\pm 16.7$ ) for the control group, with no significant difference between the groups ( $p=0.078$ ). The male-to-female ratio was 1.31 in the IgAV group and 0.99 in the control group ( $p=0.08$ ).

Skin involvement, characterised by petechiae or purpura, was significantly more common in the IgAV group (93.3%) compared with the control group (8.4%) ( $p<0.001$ ). Abdominal pain was also significantly more prevalent in the IgAV group (41%) than in the control group (12.2%) ( $p<0.001$ ). IgA deposition in biopsies was observed in 77.5% of the IgAV group, with 68.0% showing deposition in skin biopsies and 15.2% in renal biopsies, compared with significantly lower percentages in the control group ( $p<0.001$ ). In the control group, IgA deposition was observed in a total of 1.5% of biopsies, with 0.5% in skin biopsies and 1.0% in renal biopsies.

**Table 1** Demographic and clinical features of IgAV and control groups in the DCVAS cohort

| Characteristics                 | IgAV group (n=178) | Control group (n=1705) | P value |
|---------------------------------|--------------------|------------------------|---------|
| Male/female                     | 1.31               | 0.99                   | 0.08    |
| Age at onset, years, median     | 51.45 (±21.12)     | 54.09 (±16.65)         | 0.07    |
| Skin involvement                | 166 (93.3%)        | 543 (31.8%)            | <0.001  |
| Petechiae or purpura            | 131 (73.6%)        | 344 (20.2%)            | <0.001  |
| Painful skin lesion of any type | 69 (38.8%)         | 170 (10.0%)            | <0.001  |
| Maculopapular or papular rash   | 19 (10.7%)         | 144 (8.4%)             | 0.31    |
| Abdominal pain                  | 73 (41.0%)         | 208 (12.2%)            | <0.001  |
| IgA deposition in biopsy        | 138 (77.5%)        | 25 (1.5%)              | <0.001  |
| Skin biopsy                     | 121 (68.0%)        | 9 (0.5%)               | <0.001  |
| Renal biopsy                    | 27 (15.2%)         | 17 (1.0%)              | <0.001  |
| Arthritis or arthralgia         | 96 (53.9%)         | 802 (47%)              | 0.08    |
| Renal involvement               | 126 (70.8%)        | 1131 (66.3%)           | 0.23    |
| Proteinuria                     | 99 (55.6%)         | 933 (55.1%)            | 0.89    |
| Haematuria                      | 113 (63.5%)        | 982 (57.6%)            | 0.13    |
| ANCA positive                   | 15 (8.4%)          | 1320 (77.4%)           | <0.001  |
| cANCA by IF                     | 4 (2.2%)           | 671 (39.4%)            | <0.001  |
| pANCA by IF                     | 7 (3.9%)           | 460 (27.0%)            | <0.001  |
| MPO-ANCA by ELISA               | 2 (1.1%)           | 534 (31.3%)            | <0.001  |
| PR3-ANCA by ELISA               | 2 (1.1%)           | 716 (42.0%)            | <0.001  |
| Blood cryoglobulin positive     | 22 (12.4%)         | 64 (3.8%)              | <0.001  |
| ESR (mean±SD)                   | 34 (±25)           | 63 (±36)               | <0.001  |
| CRP (mean±SD)                   | 46 (±50)           | 110 (±124)             | <0.001  |

ANCA, anti-neutrophil cytoplasmic autoantibody; cANCA, cytoplasmic ANCA; CRP, C-reactive protein; DCVAS, Diagnosis and Classification in Vasculitis Study; ELISA, enzyme-linked immunosorbent assay; ESR, erythrocyte sedimentation rate; IF, immunofluorescence; IgAV, IgA vasculitis; MPO-ANCA, anti-myeloperoxidase ANCA; pANCA, perinuclear ANCA; PR3-ANCA, anti-proteinase 3 ANCA.

Arthritis or arthralgia was reported in 53.9% of the IgAV group and 47% of the control group ( $p=0.080$ ). Renal involvement, defined by proteinuria and/or haematuria, was noted in 70.8% of the IgAV group, and 66.3% in the control group ( $p=0.230$ ). The presence of ANCA and blood cryoglobulins was significantly lower in the IgAV group compared with the control group ( $p<0.001$  for all comparisons).

### Performance and revision of the Ankara 2008 IgAV classification criteria

The performance of each Ankara 2008 classification criterion for sensitivity and specificity within the DCVAS cohort is presented in [table 2](#). When the Ankara criteria were tested in the dataset, the sensitivity for classifying patients with IgAV was 91.0 (95% CI 86.9% to 95.1%) and the specificity was 73.4% (95% CI 71.3% to 75.5%) ([table 3](#)).

Since many forms of small-vessel or medium-vessel vasculitis overlap clinically with IgAV, including on the basis of skin, joint and kidney involvement, and since false positive cases meeting the original Ankara 2008 criteria in the AAV and cryoglobulinaemic vasculitis groups

decreased specificity, exclusion criteria were considered. The specificity of the criteria improved when positive tests for ANCA or blood cryoglobulins were considered as exclusion criteria: the sensitivity was 76.4% (95% CI 69.8% to 82.2%) and the specificity was 94.5% (95% CI 93.4% to 95.1%) ([table 2](#)).

### Differences between patients identified and patients not identified by the criteria

Online supplemental table 2 compares the characteristics of patients classified with IgAV by the revised Ankara 2008 criteria versus those not so classified. Patients classified by the revised criteria were more likely to be male ( $p=0.04$ ) and younger at disease onset ( $p=0.01$ ). The presence of skin involvement was significantly higher in patients meeting the revised criteria ( $p<0.001$ ), as was IgA deposition in biopsies ( $p=0.03$ ). There were no significant differences in the prevalence of abdominal pain, arthritis/arthralgia or renal involvement between the groups.

### Validation in a multicentre adult vasculitis registry

The criteria were validated in the TRVaS registry matching the eligibility criteria used for the DCVAS

**Table 2** Sensitivity and specificity of the revised Ankara 2008 criteria to classify patients as IgA vasculitis in the DCVAS cohort

| Criterion   | Sensitivity (%) | Specificity (%) | AUC (%) |
|---|-----------------|-----------------|---------|
| Skin involvement (mandatory criterion)  | 93.3            | 68.2            | 81.0    |
| (1) Abdominal pain  | 41.0            | 87.8            | 64.0    |
| (2) Histopathology*   | 77.5            | 98.5            | 88.0    |
| (3) Arthritis or arthralgias  | 54.0            | 53.0            | 53.0    |
| (4) Renal involvement   | 70.8            | 33.7            | 52.0    |
| Exclusion criteria  |                 |                 |         |
| ANCA positive   | 8.4             | 22.6            | 84.0    |
| Blood cryoglobulin positive   | 12.4            | 96.2            | 54.0    |
| Revised EULAR/PRINTO/PReS Ankara 2008 classification definition (95% CI 0.84 to 0.96) | 76.4            | 94.5            | 78.0    |

\*Histopathology confirming IgA deposition.  
ANCA, anti-neutrophil cytoplasmic autoantibody; AUC, area under curve; DCVAS, Diagnosis and Classification in Vasculitis Study; EULAR, The European Alliance of Associations for Rheumatology; PReS, Pediatric Rheumatology European Society; PRINTO, Paediatric Rheumatology International Trials Organisation.

group. The dataset consisted of 135 cases of IgAV and 133 comparators. The demographic and clinical data for both groups are presented in online supplemental table 3, with a median age at onset of 39.5 years (SD  $\pm$ 16.7). The male-to-female ratio was 1.6, indicating a slight male predominance. Skin involvement, characterised by petechiae or purpura, was observed in 98.5% of patients ( $p < 0.001$ ).

Abdominal pain was significantly more prevalent in the IgAV group (36.1%) compared with the control group (5.3%,  $p < 0.001$ ). Histopathological evidence of IgA deposition was present in 57.7% of the IgAV group ( $p < 0.001$ ). Renal involvement, defined by proteinuria or haematuria, was observed in 62.7% of IgAV patients, although this difference was not statistically significant compared with the control group ( $p = 0.24$ ).

Online supplemental table 4 compares the characteristics of patients classified with IgAV by the revised Ankara 2008 criteria versus those not so classified. Among patients meeting the criteria, all exhibited skin involvement ( $p < 0.001$ ), and abdominal pain was present in 36.9%. Histopathological confirmation of IgA deposition was observed in 57.5% of these patients.

The performance of each Ankara 2008 classification criterion for sensitivity and specificity within the DCVAS cohort is presented in online supplemental table 5. When the revised Ankara criteria were tested in the dataset, the sensitivity and specificity were 97.8% (95% CI 94.0% to 99.0%) and 85.0% (95% CI 78.0% to 90.0%), respectively.

## DISCUSSION

This study examined the performance of the original and a revised version of the EULAR/PReS-endorsed Ankara 2008 childhood classification criteria in adults with IgAV. These criteria should serve for the purpose of classification and not diagnosis of IgAV. Thus, these criteria should only be applied when other potential 'vasculitis mimics' have been excluded and a primary small-vessel vasculitis is diagnosed. The EULAR/PReS-endorsed Ankara 2008 criteria for IgAV perform well in childhood and adolescence. The SHARE (Single Hub and Access point for paediatric Rheumatology in Europe) recommendations for the diagnosis and management of vasculitis suggested these criteria be used to classify all children with IgAV. These criteria have several advantages over the 1990 ACR classification criteria:<sup>4</sup> IgA immune complexes play a crucial role in pathogenesis, whereas IgA is not included in the 1990 criteria but is a feature in the Ankara 2008 criteria. The 1990 criteria refer only to vessel wall granulocytes, which are extremely non-specific, for the histopathological criterion. One item in the ACR criteria is age  $< 20$  years, which is also misleading since age should not be a limiting factor. Finally, although palpable purpura and abdominal pain are in the ACR criteria, renal involvement and arthralgia and arthritis are not, despite being present in 33% and 78% of paediatric patients, respectively.<sup>3</sup> These findings align with prior work indicating that IgAV may present differently across age groups,<sup>13,14</sup> supporting the development of classification criteria specifically for adults.

The sensitivity and specificity of the Ankara 2008 criteria are 100% and 87%, respectively, in the childhood series.<sup>4</sup> Hočevar *et al* have shown that Ankara 2008 criteria had 99.2% (95% CI 95.4% to 99.9%) sensitivity and 86.0% (95% CI 80.7% to 90.3%) specificity in adult IgAV patients.<sup>15</sup> On the other hand, the specificity was low when the original criteria were applied directly to adult IgAV patients in the DCVAS registry. This was mainly due to the presence of cryoglobulinemic vasculitis in adults and to the overlapping features of adult IgAV with AAV. Thus, an exclusion criterion was added to classify adults, with a positive test for ANCA or blood cryoglobulins serving as exclusion factors. To apply the same criteria throughout all age groups, the same exclusion criteria can be applied for children as a revised Ankara 2008 criterion. This is not expected to change the sensitivity and specificity in children, which is already high. Furthermore, cryoglobulinemic vasculitis is extremely rarely observed in children.

IgAV is the most common vasculitis in childhood. Paediatricians need reliable criteria to classify patients. The development of the Ankara 2008 criteria reflected the collaborative effort of international experts in paediatric vasculitis, including rheumatologists and nephrologists. The criteria define the important clinical features that differentiate IgAV from other types of small-vessel and medium-vessel vasculitis. The registry used for the development and validation of the criteria included patients

**Table 3** Sensitivity and specificity of Ankara 2008 criteria in DCVAS, TRVaS and the original paediatric cohort

| Criteria set   | Glossary  | Sensitivity (%) | Specificity (%) | AUC (%) |
|--|---|-----------------|-----------------|---------|
| EULAR/PRINTO/PReS Ankara 2008 classification definition in the original paediatric cohort: | Skin involvement and at least one of the four following criteria:<br>1. Abdominal pain<br>2. Histopathology<br>3. Arthritis or arthralgia<br>4. Renal involvement   | 100.0           | 87.0            | 93.5    |
| DCVAS cohort: Original EULAR/PRINTO/PReS Ankara 2008 criteria:                             | Skin involvement and at least one of the four following criteria:<br>1. Abdominal pain<br>2. Histopathology<br>3. Arthritis or arthralgia<br>4. Renal involvement   | 91.0            | 73.4            | 63.0    |
| DCVAS cohort: Revised EULAR/PRINTO/PReS Ankara 2008 criteria:                              | Skin involvement and at least one of the four following criteria:<br>1. Abdominal pain<br>2. Histopathology<br>3. Arthritis or arthralgia<br>4. Renal involvement<br>Exclusion criteria:<br>ANCA and/or blood cryoglobulin positivity | 76.4            | 94.5            | 78.0    |
| TRVaS validation cohort: Revised EULAR/PRINTO/PReS Ankara 2008 criteria                    | Skin involvement and at least one of the four following criteria:<br>1. Abdominal pain<br>2. Histopathology<br>3. Arthritis or arthralgia<br>4. Renal involvement<br>Exclusion criteria:<br>ANCA and/or blood cryoglobulin positivity | 97.8            | 85.0            | 91.0    |

ANCA, anti-neutrophil cytoplasmic autoantibody; AUC, area under curve; DCVAS, Diagnosis and Classification in Vasculitis Study; EULAR, European Alliance of Associations for Rheumatology; PReS, Pediatric Rheumatology European Society; PRINTO, Paediatric Rheumatology International Trials Organisation; TRVaS, Turkish Vasculitis Study Group.

from various parts of the world. Classification criteria are needed to define patients for collaborative studies and to harmonise relevant data for the disease.

There are several strengths to the current study. The DCVAS database is derived from a multinational study, in contrast to the 1990 criteria, which only included adult patients from North America. The comparator group for validating the new criteria mainly included other small-vessel vasculitides that may mimic IgAV, where differentiation from IgAV may be challenging but important. The original criteria for IgAV were developed by a large group of international experts, validated in children, and then endorsed by EULAR. Finally, these revised criteria for adults were validated in an independent (Turkish) cohort.

There are also limitations in the current study to consider. For validation, only patients whose diagnostic certainty was >75% were included; however, the diagnosis relied entirely on the judgement of the registering physician. 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 under-represented in the DCVAS cohort and may have different clinical presentations of IgAV. Other limitations include

a lack of consistency in the pathology definitions and the possibility of missing data.

Adopting the proposed revised Ankara criteria for IgAV in adults, and using these criteria along with the original Ankara criteria for children, will enable studies across the lifespan to further explore and understand the pathogenesis and best management of IgAV.

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