

Supplementary Table S1. Positivity of 2022 ACR/EULAR criteria items for MPA.

	MPA, n=355	GPA, n=99	EGPA, n=104
Nasal involvement, n (%)	6 (1.7)	26 (26.3)	7 (6.7)
MPO-ANCA positive, n (%)	342 (96.3)	57 (57.6)	36 (34.6)
Interstitial pneumonia, n (%)	191 (53.8)	19 (19.2)	15 (14.4)
Pauci-immune glomerulonephritis, n (%)	136 (38.3)	24 (24.2)	2 (1.9)
PR3-ANCA positive, n (%)	10 (2.8)	39 (39.4)	3 (2.9)
Blood eosinophil count $\geq 1 \times 10^9$ /litre, n (%)	9 (2.5)	2 (2.0)	92 (88.5)

Positivity was analysed for each of the items in the 2022 ACR/EULAR criteria for patients diagnosed as MPA in the J-CANVAS registry.

ANCA: anti-neutrophil cytoplasmic antibody; EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; J-CANVAS: Japan Collaborative registry of ANCA-associated VASculitis; MPA: microscopic polyangiitis; MPO: myeloperoxidase; PR3: proteinase 3.

Supplementary Table S2. Positivity of 2022 ACR/EULAR criteria items for GPA.

	MPA, n=355	GPA, n=99	EGPA, n=104
Nasal involvement, n (%)	6 (1.7)	26 (26.3)	7 (6.7)
Cartilaginous involvement, n (%)	0 (0.0)	1 (1.0)	1 (0.96)
Hearing loss, n (%)	30 (8.5)	33 (33.3)	1 (0.96)
PR3-ANCA positive, n (%)	10 (2.8)	39 (39.4)	3 (2.9)
Granulomatous lesion, n (%)	163 (46.0)	62 (62.6)	34 (32.7)
Rhinosinusitis, n (%)	25 (7.0)	36 (36.4)	37 (35.6)
Pauci-immune glomerulonephritis, n (%)	136 (38.3)	24 (24.2)	2 (1.9)
Pulmonary nodule, n (%)	21 (5.9)	40 (40.4)	8 (7.7)
MPO-ANCA positive, n (%)	342 (96.3)	57 (57.6)	36 (34.6)
Blood eosinophil count $\geq 1 \times 10^9$ /litre, n (%)	9 (2.5)	2 (2.0)	92 (88.5)

Positivity was analysed for each of the items in the 2022 ACR/EULAR criteria for patients diagnosed as GPA in the J-CANVAS registry.

ANCA: anti-neutrophil cytoplasmic antibody; EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; J-CANVAS: Japan Collaborative registry of ANCA-associated VASculitis; MPA: microscopic polyangiitis; MPO: myeloperoxidase; PR3: proteinase 3.

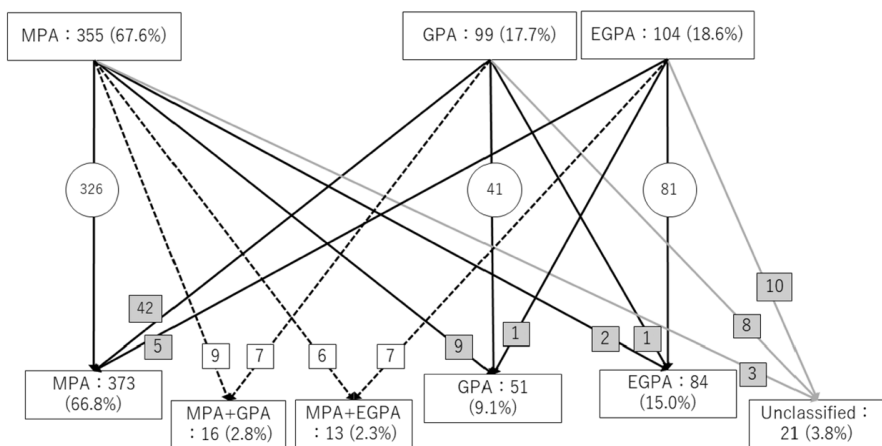
Supplementary Table S3. Positivity of 2022 ACR/EULAR criteria items for EGPA.

	MPA, n=355	GPA, n=99	EGPA, n=104
Obstructive airway disease, n (%)	21 (5.9)	8 (8.1)	92 (88.5)
Nasal polyps, n (%)	6 (1.7)	26 (26.3)	7 (6.7)
Mononeuritis multiplex, n (%)	34 (9.6)	4 (4.0)	55 (52.9)
Blood eosinophil count $\geq 1 \times 10^9$ /litre, n (%)	9 (2.5)	2 (2.0)	92 (88.5)
Extravascular Eosinophilic-predominant inflammation on biopsy, n (%)	163 (45.9)	62 (62.6)	34 (32.7)
PR3-ANCA positive, n (%)	10 (2.8)	39 (39.4)	3 (2.9)
Haematuria, n (%)	249 (70.1)	51 (51.5)	24 (23.1)

Positivity was analysed for each of the items in the 2022 ACR/EULAR criteria for patients diagnosed as EGPA in the J-CANVAS registry.

ANCA: anti-neutrophil cytoplasmic antibody; EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; J-CANVAS: Japan Collaborative registry of ANCA-associated VASculitis; MPA: microscopic polyangiitis; PR3: proteinase 3.

J-CANVAS Diagnosis → 2022 ACR/EULAR criteria

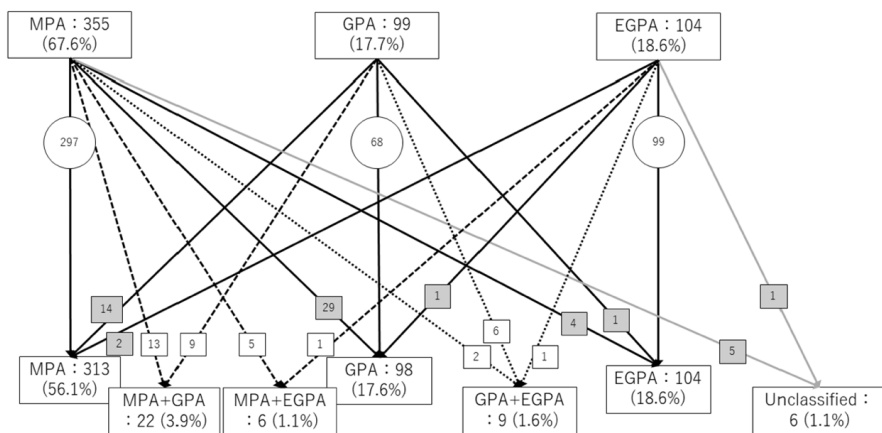


Supplementary Fig. S1. Reclassification from J-CANVAS Diagnosis to 2022 ACR/EULAR Criteria.

J-CANVAS diagnosis: ANCA associated vasculitis diagnosis was based on the 2012 CHCC definition and European Medicines Agency algorithm. The diagnosis was ultimately made by expert rheumatologists, nephrologists, or pulmonologists.

EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis.

J-CANVAS Diagnosis → modified criteria



Supplementary Fig. S2. Reclassification from J-CANVAS diagnosis to modified criteria.

J-CANVAS diagnosis: ANCA associated vasculitis diagnosis was based on the 2012 CHCC definition and European Medicines Agency algorithm. The diagnosis was ultimately made by expert rheumatologists, nephrologists, or pulmonologists.

EGPA: eosinophilic granulomatosis with polyangiitis; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis.

Modified classification criteria for Japanese AAV patients
MICROSCOPIC POLYANGIITIS

CONSIDERATIONS PRIOR TO APPLICATION

- This criteria should be applied to patients diagnosed with ANCA-associated vasculitis.

Nasal involvement	-2.3
Hearing loss	-1.5
Rhinosinusitis	-1.3
Obstructive airway disease	-1.8
MPO-ANCA positive	+2.7
Blood eosinophil count $\geq 1 \times 10^9/\text{liter}$	-3.0
Hematuria	+0.8
Fibrosis or interstitial lung disease on chest imaging	+1.6
Pulmonary nodules on chest imaging	-2.3
If the sum of the scores for 9 items are ≥ 1.23 , the patient is classified as MICROSCOPIC POLYANGIITIS.	

Supplementary Fig. S3. The modified classification criteria for microscopic polyangiitis.

Each item score was derived from the regression coefficients identified in the multivariable analysis, and a cut-off value of 1.23 or higher was used to classify patients as microscopic polyangiitis.

Modified classification criteria for Japanese AAV patients
GRANULOMATOSIS WITH POLYANGIITIS

CONSIDERATIONS PRIOR TO APPLICATION

- This criteria should be applied to patients diagnosed with ANCA-associated vasculitis.

Nasal involvement	+2.5
Hearing Loss	+1.9
Rhinosinusitis	+0.9
Mononeuritis multiplex	-1.8
PR3-ANCA positive	+3.0
Blood eosinophil count $\geq 1 \times 10^9$ /liter	-3.0
Pulmonary nodules on chest imaging	+2.5
Fibrosis or interstitial lung disease on chest imaging	-1.4

If the sum of the scores for 8 items are ≥ 1.22 the patient is classified as GRANULOMATOSIS WITH POLYANGIITIS.

Supplementary Fig. S4. The modified classification criteria for granulomatosis with polyangiitis.

Each item score was derived from the regression coefficients identified in the multivariable analysis, and a cut-off value of 1.22 or higher was used to classify patients as granulomatosis with polyangiitis.

Modified classification criteria for Japanese AAV patients
EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS

CONSIDERATIONS PRIOR TO APPLICATION

- This criteria should be applied to patients diagnosed with ANCA-associated vasculitis.

Mononeuritis multiplex	+2.0
Obstructive airway disease	+4.0
Pauci-immune glomerulonephritis	-2.5
MPO-ANCA positive	-1.4
Blood eosinophil count $\geq 1 \times 10^9$ /liter	+5.0
Fibrosis or interstitial lung disease on chest imaging	-1.9

If the sum of the scores for 6 items are ≥ 1.85 , the patient is classified as EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS.

Supplementary Fig. S5. The modified classification criteria for eosinophilic granulomatosis with polyangiitis.

Each item score was derived from the regression coefficients identified in the multivariable analysis, and a cut-off value of 1.85 or higher was used to classify patients as eosinophilic granulomatosis with polyangiitis.