

Medical or Research Professionals/Clinicians

Topic area: Clinical topics by disease

Topic: 17. Vasculitis

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DRAFT CLASSIFICATION CRITERIA FOR THE ANCA ASSOCIATED VASCULITIDES

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My abstract has been or will be presented at a scientific meeting during a 12 months period prior to EULAR 2018: No
Is the first author applying for a travel bursary and/or an award for undergraduate medical students?: No

Background: Classification criteria for the ANCA-associated vasculitides (AAVs) were developed in the 1980s prior to the use of ANCA testing and newer imaging techniques. The Diagnostic and Classification of the Systemic Vasculitides (DCVAS) study is an international project to update classification criteria for the systemic vasculitides.

Objectives: Development of draft classification criteria for Granulomatosis with Polyangiitis (GPA), Microscopic Polyangiitis (MPA) and Eosinophilic Granulomatosis with Polyangiitis (EGPA).

Methods: Three phases: 1) Expert panel review of cases to identify gold standard set of new cases of small vessel vasculitis; 2) Item reduction of >8000 individual DCVAS items using data-driven and consensus methodology; 3) Lasso logistic regression models within each development set comparing each of the AAV types to other small and medium vessel vasculitides. Final criteria derived through clinical consensus, tested in validation set. The classification project has received financial support from the ACR and EULAR.

Results: The expert review process approved 2072/2871 (72%) of physician diagnosed DCVAS cases, including [724 GPA, 291 MPA, 226 EGPA, 51 polyarteritis nodosa (PAN), 220 other small vessel disease (SVV)]. Data driven and expert consensus resulted in 91 items retained. Draft criteria, and sensitivity and specificity in Table 1.

Granulomatosis with polyangiitis (GPA)	Microscopic polyangiitis (MPA)	Eosinophilic granulomatosis with polyangiitis (EGPA)
Blood nasal discharge, ulcers, crusting, congestion or blockage, or septal defect/perforation +3	Pauci-immune glomerulonephritis +3	Obstructive airways diseases +3
Cartilagenous involvement*+2	Bloody nasal discharge, ulcers, crusting, congestion or blockage, septal defect/ perforation -3	Nasal polyps +3
Conductive or sensorineural hearing loss +1	pANCA or MPO-antibody positive +6	Mononeuritis multiplex or motor neuropathy +1
Pauci-immune glomerulonephritis +1	Fibrosis or ILD on chest imaging +3	Eosinophil count $\geq 1 \times 10^9/L$ +5
cANCA or PR3-antibody +5	cANCA or PR3-antibody -1	Extravascular eosinophilic predominant inflammation/ eosinophils in bone marrow +2
pANCA or MPO-antibody -1	Eosinophil count $\geq 1 \times 10^9/L$ -4	cANCA or PR3-antibody -3
Eosinophil count $\geq 1 \times 10^9/L$ -4		Microscopic haematuria -1
Granuloma, extravascular granulomatous inflammation, or giant cells on biopsy +2		
Nodules, mass, cavitation on chest imaging +2		
Inflammation, consolidation, or effusion of the nasal/paranasal sinuses on imaging +1		

Total score of ≥ 5 is needed for classification Sensitivity 93%, Specificity 94%	Total score of ≥ 6 is needed for classification Sensitivity 87%, Specificity 96%)	Total score of ≥ 5 is needed for classification Sensitivity 88%, Specificity 98%
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Table 1. Draft classification criteria for the ANCA-associated vasculitides. *Cartilagenous involvement: Inflamed ear or nose cartilage or hoarse voice/stridor, endobronchial involvement or saddle nose deformity

Conclusions: Draft classification criteria for GPA, MPA and EGPA have been created which reflect current practice and have good sensitivity and specificity.

References:

Acknowledgements: DCVAS sites and expert panel members

Disclosure of Interest: None declared