

## Towards diagnostic criteria for the ANCA-associated vasculitides

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The primary systemic vasculitides are an important cause of mortality and morbidity in the community. Their incidence is >100 new cases per million per year (1). There are significant benefit with intensive chemotherapy but if the diagnosis is delayed, this may adversely affect the outcome. Up to 25% of patients on chronic haemodialysis programmes may have had undiagnosed systemic vasculitis as a major contributor to renal failure (2).

Classification criteria are useful to confirm that a group of patients with a clinical diagnosis have a similar or identical condition. However, in order to discriminate between patients with or without a specific disease, diagnostic criteria are required. There are currently no validated diagnostic criteria for primary systemic vasculitis although there are classification criteria that have been misapplied – the American College of Rheumatology (ACR) criteria (3) and the Chapel Hill Consensus Conference (CHCC) Definitions (4). When tested, the CHCC definitions supplemented with surrogate clinical and laboratory parameters failed to act as diagnostic criteria (5). Diagnostic criteria for Behçet's disease, which is classified as a secondary vasculitis, have been developed for use in clinical trials (6).

Diagnostic criteria for primary systemic vasculitis would be most useful for practising clinicians who could rapidly and accurately assess the probability of vasculitis in the context of patients who present to them acutely, especially those with multi-system ill-

nesses. There are currently no satisfactory serological tests to enable a rapid diagnosis of vasculitis. The ANCA test, when used by non-specialists has a diagnostic yield of only 0-12% for systemic vasculitis in a routine clinical setting (7). At present, ANCA testing is not part of the existing classification criteria for vasculitis. The considerable overlap of disease features across the ANCA related vasculitides is further justification to re-evaluate diagnostic and classification criteria (8).

### References

1. GONZALEZ-GAY MA, GARCIA-PORRUA C: Systemic vasculitis in adults in Northwestern Spain, 1988-1997: Clinical and epidemiologic aspects. *Medicine* 1999; 78: 292-308
2. ADU D, HOWIE AJ, SCOTT DG *et al.*: Polyarteritis and the kidney. *Q J Med* 1987; 62: 221-37.
3. HUNDER HH, AREND WP, BLOCH DA *et al.*: The American college of Rheumatology 1990 criteria for the classification of vasculitis: introduction. *Arthritis Rheum* 1990; 33: 1066-7.
4. JENNETTE JC, FALK RJ, ANDRASSY K *et al.*: Nomenclature of systemic vasculitides: proposal of an international consensus conference. *Arthritis Rheum* 1994; 37: 187-92.
5. SORENSEN SF, SLOT O, TVEDE N *et al.*: A prospective study of vasculitis patients collected in a five year period: evaluation of the Chapel Hill nomenclature. *Ann Rheum Dis* 2000; 59: 478-82.
6. INTERNATIONAL STUDY GROUP FOR BEHÇET'S DISEASE: Criteria for diagnosis of Behçet's disease. *Lancet* 1990; 336: 1078-80.
7. MCLAREN JS, STIMSON RH, MCRORIE ER *et al.*: The diagnostic value of anti-neutrophil cytoplasmic antibody testing in a routine clinical setting. *Q J Med* 2001; 94: 615-21.
8. JENNETTE JC, FALK RJ: Nosology of primary vasculitis. *Curr Opin Rheumatol* 2007; 19: 10-16.

**Table I.** Limitations of current definitions and classification of ANCA associated vasculitis.

Disease	Current classification(s)	Problems
Wegener's granulomatosis	CHCC definition – Small vessel vasculitis ACR classification – Wegener's granulomatosis	Overlap with MPA; range of organ involvement Some aspects are not vasculitic (eg nasal granulomata)
Microscopic polyangiitis	CHCC definition – Small vessel vasculitis ACR classification – no definition	Renal only variety may evolve into WG or MPA
Churg Strauss Syndrome	CHCC definition – Small vessel vasculitis ACR classification – Churg Strauss syndrome	Overlap with WG; renal disease less common than for WG and MPA; some aspects are non-vasculitic (eg allergies)
Unspecified small vessel vasculitis	CHCC definition – Unspecified small vessel vasculitis ACR classification – Unspecified small vessel vasculitis	May overlap features of MPA/WG/CSS or be more limited; less well studied; some patients remain unclassifiable and should not be "forced" into an artificial system to describe the condition