Evolution of paediatric-specific vasculitis classification criteria

Sarah Ringold, Carol A Wallace

This issue of *Annals of the Rheumatic Diseases* includes two articles describing the proposed paediatric-specific classification criteria for childhood polyarteritis nodosa (c-PAN), Wegener granulomatosis (c-WG), Takayasu arteritis (c-TA) and Henoch–Schönlein purpura (HSP) which resulted from an ambitious multicentre, international collaborative project that was initiated in Vienna in 2005 and culminated in the 2008 Ankara Consensus Conference (see articles on pages 790 and 798).1 2

The vasculitides are a heterogeneous collection of multisystem disorders that manifest as the inflammation of blood vessels and may occur as a primary process or may be secondary to another disorder, most commonly infection. Despite this commonality, the vasculitides differ significantly in their clinical, radiographic, laboratory and histopathological features. Much of this heterogeneity arises from the different sizes of the blood vessels that are primarily affected, the involvement of arteries, veins, or both, and from the varying patterns of organ involvement, all of which have important implications for the optimal medical treatment and prognosis of the disorders. Perhaps some of the most interesting and significant variations within the vasculitides are those associated with the age of onset. For example, the prevalence of certain vasculitides varies considerably by age of onset. Kawasaki disease (KD), one of the most common paediatric vasculitides, only rarely manifests in adulthood, while other vasculitides, such as giant cell arteritis, are seen only in adulthood. Other vasculitides have different patterns of organ involvement and differing prognoses depending on age of onset, such as in HSP, for which the prevalence of arthritis and the prognosis associated with kidney involvement both vary significantly according to the age of onset.3 4

Development of classification criteria for the vasculitides has been particularly challenging given these variable disease presentations and the evolution of laboratory and radiographic techniques that have changed the way in which the vasculitides are evaluated and diagnosed. Some of the most significant developments have been the development of the antinuclear cytoplasmic antibody (ANCA) assay and the increased use of techniques such as CT angiography and magnetic resonance angiography, which are increasingly used in diagnosing these disorders and in some cases are used in place of tissue biopsy.

In general, the goal of a set of classification criteria is to identify relatively homogeneous, non-overlapping groups of patients who share a common disorder, and they are generally defined as a group of clinical or other findings that predict the presence of the defining aspects of the disease.5 Diagnostic criteria, on the other hand, are derived to help clinicians distinguish vasculitis from non-vasculitis in their daily practice. In the case of the vasculitides, the classification criteria are based upon clinical, laboratory, histopathological and radiographic characteristics and are intended to differentiate patients who have been diagnosed with vasculitis into specific subtypes. The two sets of criteria therefore serve distinct purposes. Sensitive and specific classification criteria remain of key importance for defining cohorts for study in epidemiological, basic science and clinical research.

The classification criteria for vasculitides have undergone numerous iterations which reflect improvements in recognition and diagnosis of the disorders and the increased availability of published clinical reports describing case characteristics and outcomes. One of the initial significant developments in the study of vasculitis was the first formal scheme for the classification of vasculitides that was published in 1952 by Zeek.6 The criteria required the presence of necrotising angitis and were based primarily on the size of the vessels affected and the pattern of organ involvement. The criteria defined classification criteria for five different types of vasculitis: (1) hypersensitivity angitis; (2) allergic granulomatous angitis; (3) rheumatic arteritis; (4) periarteritis nodosa; (5) temporal arteritis. The emphasis on vessel size led the way for the development of additional sets of classification criteria and modifications centred upon this defining characteristic.

In the 1980s the American College of Rheumatology (ACR) established a subcommittee to derive classification criteria for seven specific categories of vasculitis: giant cell arteritis, TA, WG, Churg–Strauss syndrome (CSS), PAN, HSP and hypersensitivity vasculitis. These criteria, published in 1990, were derived from a prospectively developed cohort of 1000 patients with vasculitis from 46 centres in Mexico, Canada and the USA and provided classification criteria in both traditional and tree classification formats.7 The resultant criteria had sensitivities and specificities, ranging from 70% to 99%. However, these criteria were developed before the widespread use of ANCA and did not include classification criteria for microscopic polyangiitis (MPA), which was being increasingly described in the literature, but had not yet been formally recognised as a separate entity.

Given concerns about the continued lack of standardised nomenclature and disease definitions, the Chapel Hill Consensus Conference on the Nomenclature of Systemic Vasculitis was convened in 1994 and included an international, multidisciplinary committee which used modified nominal group techniques to propose a set of definitions for the vasculitides (ie, the elements of each disease that are present in every case of the disease).8 The primary contributions of this conference were to include a definition for MPA and to acknowledge that, owing to the limitations in histological data such as the typically patchy involvement which may result in the biopsy not being representative, in some cases surrogate markers could be acceptable in place of tissue. Subsequent efforts have tried to derive classification criteria from these definitions with varying success.9 10

It was recognised early on that these classification criteria, derived from adult cohorts, would not have acceptable sensitivity and/or specificity for use in paediatrics. In particular, the lack of classification criteria for KD and HSP, two of the most common paediatric vasculitides, and the lack of inclusion of ANCA in the criteria were felt to be significant limitations. In response to these concerns, the Pediatric Rheumatology European Society along with the European League Against Rheumatism established...
a working group consisting of paediatric rheumatologists and nephrologists whose objective was to assess whether the 1990 ACR criteria were adequate for paediatrics, and, if not, to develop a pre-
liminary set of paediatric-specific clas-
ification criteria that would adequately reflect the differences between paediatric
and adult disease and would incorporate new diagnostic tools. This conference
was held in Vienna in 2005 and, after a
review of the literature and expert con-
sensus, as derived from a combination of
Delphi and nominal group techniques, it
was decided to base the criteria on ves-
sel size, similar to the 1990 ACR criteria,
and the small-vascular vasculitis subgroup
was further divided into granuloma-
tous and non-granulomatous vasculitis.
The ‘Other vasculitides’ category was
expanded to include such diagnoses as
Behçet disease and Cogan syndrome.

Preliminary criteria were subsequently
proposed for c-PAN, WG, HSP, TA and
KD and the planning for data collection
for the prospective validation of these
criteria was begun. The final examina-
tion of the data and consensus processes,
as described in this issue of the Annals,
subsequently occurred in 2008 at the
Ankara Consensus Conference.

Some of the major changes proposed at
the 2005 Vienna conference included the
removal of the age criterion from the HSP
criteria and the inclusion of the presence
of IgA deposition on biopsy as a criterion.
Palpable purpura was added as a manda-
tory criterion. Serological evidence of
streptococcal infection was added to the
criteria for PAN, recognising the frequent
association between streptococcal infec-
tion and PAN in children. The WG crite-
reria were augmented to include subglottic,
endotracheal and/or endobronchial ste-
ria were augmented to include subglottic,
association between streptococcal infec-
tion was added to the criteria and coronary artery
stenosis was added as a mandatory cri-
terion. Instead of plain radiographs was also
used fluorescence. The use of a CT scan
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Ann Rheum Dis 2010 69: 785-786 originally published online April 13, 2010
doi: 10.1136/ard.2009.127886

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