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Update in paediatric vasculitis

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Vasculitis refers to a heterogeneous group of disorders that are characterised by inflammatory destruction of blood vessels. Although simple to define, almost nothing about vasculitis is simple. From classification to diagnosis, and from pathogenesis to management, large gaps remain in our understanding. Despite extensive and ongoing research, the fundamental mechanisms underlying the initiation and continuation of systemic vasculitis remain poorly understood. Thus, vasculitis continues to provide tremendous challenges to both clinicians and investigators and remains a rich source of issues for discussion. This review concentrates on recent changes proposed for the classification of paediatric vasculitis and advances in the concepts of aetiopathogenesis. Availability of improved classification criteria for children should prompt planning for multicentre-controlled studies for the treatment of these rare but important diseases.

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New concepts in classification

The classification of vasculitides has been controversial since the first attempts more than a century ago due, in large part, to a general lack of understanding regarding the mechanisms of disease

Abbreviations: American College of Rheumatology, ACR; Anti-neutrophil cytoplasmic antibodies, ANCA; C-reactive protein, CRP; Churg–Strauss syndrome, CSS; European League Against Rheumatism, EULAR; European Vasculitis Study, EUVAS; Henoch–Schonlein purpura, HSP; Intravenous immunoglobulin, IVIG; Kawasaki disease, KD; Microscopic polyangiitis, MPA; Myeloperoxidase, MPO; Paediatric Rheumatology European Society, PRES; Polyarteritis nodosa, PAN; Primary angiitis of the central nervous system, PACNS; Proteinase-3, PR3; Pulse intravenous methylprednisolone, IVMP; Takayasu arteritis, TA; Tumour necrosis factor, TNF; Wegener granulomatosis, WG.

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pathogenesis [1]. Most current classification systems are based on a combination of histological and clinical features. A consensus committee convened by the American College of Rheumatology (ACR) in 1990 proposed classification criteria based on adult data [2]. The ACR Vasculitis Study Committee, however, limited itself to defining 'primary' vasculitis characteristics and only considered seven types of vasculitis. These, and the classification systems proposed by Lie [3] and by the Chapel Hill consensus conference [4] in 1994, have formed the basis of current practice. These systems are not designed for diagnosis of vasculitis, but as entry criteria for research studies. They omit important clinical forms of vasculitis and are, in general, inadequate for clinical application. The fact they were developed based on adult data alone is also a drawback for paediatric practice.

As new data emerge in vasculitis, as in other complex medical syndromes, conditions previously thought to be similar turn out to differ in fundamental ways and conditions previously thought to be separate are found to share pathogenic mechanisms. For example, polyarteritis nodosa (PAN) classically refers to a medium-sized muscular arteritis. Although most cases involve both visceral and cutaneous vessels, disease limited to skin (cutaneous PAN), or involving the eyes and inner ears (Cogan's syndrome), have been described [5]. The identification of PAN patients with anti-neutrophil cytoplasmic antibodies (ANCAs) to myeloperoxidase (MPO) has led to the addition of ANCA-positive microscopic PAN to the list of variants [6]. The result is that use of the term PAN today refers to a broad array of conditions, some of which are limited – others systemic, some benign and others life-threatening.

Clearly, any classification system will require modification as knowledge advances. Unfortunately the tools for doing so in a coherent manner for vasculitis have remained elusive. An International Consensus Conference, formed with the support of European League against Rheumatism (EULAR) and the Pediatric Rheumatology European Society (PRES), met in 2006 and proposed the first true paediatric classification scheme (Table 1) [7]. Using standard consensus techniques, the classification committee tried to incorporate recent advances in diagnostic imaging and current knowledge regarding pathogenesis of specific disorders. For example, the presence of IgA immune complexes in biopsy material has been suggested as a part of the criteria for Henoch–Schonlein purpura (HSP), and ANCA have been included as a diagnostic feature of WG. The proposed criteria for HSP, PAN and Wegener granulomatosis (WG) have been validated and updated by an international consensus

Table 1
Classification of childhood vasculitis (Adapted from EULAR/PReS endorsed criteria for the classification of childhood vasculitides (7)).

I	Predominantly large-vessel vasculitis • Takayasu arteritis
II	Predominantly medium-sized vessel vasculitis • Childhood polyarteritis nodosa • Cutaneous polyarteritis • Kawasaki disease
III	Predominantly small vessels vasculitis (A) GRANULOMATOUS • Wegener's granulomatosis • Churg–Strauss syndrome (B) NON-GRANULOMATOUS • Microscopic polyangiitis • Henoch–Schönlein purpura • Isolated cutaneous leukocytoclastic vasculitis • Hypocomplementemic urticarial vasculitis
IV	Other vasculitides • Behçet disease • Vasculitis secondary to infection (including hepatitis B associated polyarteritis nodosa), malignancies, and drugs, including hypersensitivity vasculitis • Vasculitis associated with connective tissue diseases • Isolated vasculitis of the central nervous system • Cogan syndrome • Unclassified

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