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Systemic Vasculitis

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DEFINITION AND EPIDEMIOLOGY

The primary systemic vasculitides are inflammatory disorders of blood vessels that are characterized by immune-mediated injury leading to vessel necrosis, thrombosis, stenosis, or some combination of these. Vessels in any organ may be affected, but each vasculitis is characterized by different preferential vessel size or territory and tissue targeting. Although these disorders are rare, they may be organ- or life-threatening, so prompt diagnosis and treatment are necessary. The vasculitides are defined according to the 1990 American College of Rheumatology (ACR) classification criteria and the 1994 Chapel Hill Consensus Conference (CHCC) based on generally affected vessel size (small, medium, or large). Antineutrophil cytoplasmic antibody (ANCA)–associated vasculitides (AAVs) have known associations with characteristic autoantibodies. Figure 81-1 shows the major types of vasculitides. Although the ACR and CHCC definitions were not designed as diagnostic criteria, classification criteria such as these are important in clinical research study design, treatment, and prognosis. The ACR and the European League Against Rheumatism (EULAR) are currently in the process of refining diagnostic and classification criteria for primary vasculitides.

Determining the incidence and prevalence of each of the vasculitides is challenging given the rarity of the disorders, imperfect classification criteria and definitions for epidemiologic purposes, and some clinicopathologic overlaps that occur between certain types (e.g., AAVs).

Small Vessel Vasculitis

ANCA-Associated Vasculitides

Granulomatosis with polyangiitis (GPA; previously known as Wegener's granulomatosis), microscopic polyangiitis (MPA), eosinophilic granulomatosis with polyangiitis (EGPA; previously known as Churg-Strauss syndrome), and renal-limited vasculitis (RLV) affect small and medium-sized blood vessels and may be associated with ANCA. Various studies have shown AAVs to have an incidence of approximately 10 to 20 per million. The peak age at onset is 65 to 74 years, with a female-to-male ratio of 1.5 : 1. EGPA is the least common of the AAVs, with an incidence of approximately 1.0 to 3.0 per million, and it also has a weaker association with ANCA than GPA and MPA do.

Henoch-Schönlein Purpura

Henoch-Schönlein purpura (HSP) is a small vessel vasculitis that occurs most frequently in young children, with a peak age at onset of 4 to 6 years, but can also occur in adults. HSP accounts for almost half of all cases of childhood vasculitis. In children

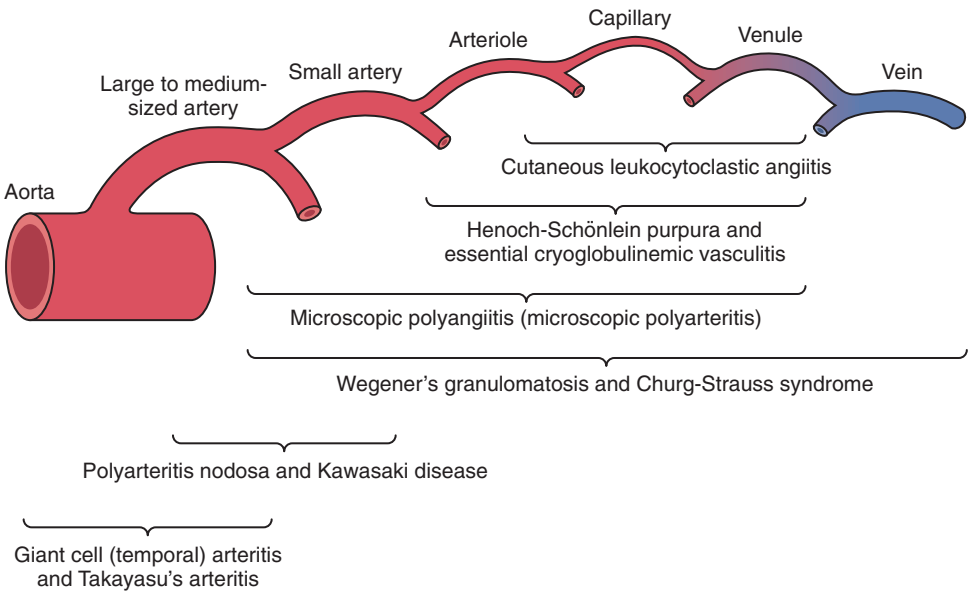

FIGURE 81-1 The vascular spectrum of the vasculitides. (From Jennette JC, Falk RJ, Andrassy K, et al.: Nomenclature of systemic vasculitides: proposal of an international consensus conference, Arthritis Rheum 37:187–192, 1994.)