

Names and definitions of vasculitides adopted by the Chapel Hill Consensus Conference on the Nomenclature of Systemic vasculitis *

Large vessel vasculitis

- Giant cell (temporal) arteritis

Granulomatous arteritis of the aorta and its major branches, with a predilection for the extra cranial branches of the carotid artery. *Often involves the temporal artery. Usually occurs in patients older than 50 and often is associated with polymyalgia rheumatica.*

- Takayasu's arteritis

Granulomatous inflammation of the aorta and its major branches. *Usually occurs in patients younger than 50.*

Medium sized vessel vasculitis

- Polyarteritis nodosa† (classic polyarteritis nodosa)

Necrotizing inflammation of medium-sized or small arteries without glomerulonephritis or vasculitis in arterioles, capillaries, or venules.

- Kawasaki disease

Arteritis involving large, medium sized, small arteries, and associated with mucocutaneous lymph node syndrome. *Coronary arteries are often involved. Aorta and veins may be involved. Usually occurs in children.*

Small vessel vasculitis

- Wegener's granulomatosis‡

Granulomatous inflammation involving the respiratory tract, and necrotizing vasculitis affecting small to medium-sized vessels (e.g. capillaries, venules, arterioles, and arteries). *Necrotizing glomerulonephritis is common.*

- Churg Strauss syndrome‡

Eosinophil-rich and granulomatous inflammation involving the respiratory tract, necrotizing vasculitis affecting small to medium-sized vessels, and associated with asthma and eosinophilia.

- Microscopic polyangiitis† (microscopic polyarteritis)‡ Necrotizing vasculitis, with few or no immune deposits, affecting small vessels (i.e. capillaries, venules, or arterioles). *Necrotizing arteritis involving small and medium sized arteries may be present. Necrotizing glomerulonephritis is very common. Pulmonary capillaritis often occurs.*

- Henoch Schönlein Purpura

Vasculitis, with IgA-dominant immune deposits, affecting small vessels i.e. capillaries, venules, or arterioles). *Typically involves skin, gut, and glomeruli, and is associated arthralgia or arthritis.*

Classification of vasculitis

- Essential cryoglobulinaemic vasculitis

Vasculitis, with cryoglobulin immune deposits, affecting small vessels (i.e. capillaries, venules, or arterioles), and associated with cryoglobulins in serum. *Skin and glomeruli are often involved.*

- Cutaneous leucocytoclastic angiitis

Isolated cutaneous leucocytoclastic angiitis without systemic vasculitis glomerulonephritis.

* Large vessel refers to the aorta and the largest branches directed towards the major body regions (e.g. to the extremities and the head and neck); medium sized vessel refers to the main visceral arteries (e.g. renal, hepatic, coronary, and mesenteric arteries); small vessel refers to venules, capillaries, arterioles, and the intraparenchymal distal arterial radicals that connect with arterioles. Some small and large vessel vasculitides may involve medium-sized arteries, but large and medium sized vessel vasculitides do not involve smaller than arteries. essential components are represented by normal type; italicised type represents usual, but not essential, components.

†Preferred term.

‡Strongly associated with antineutrophil cytoplasmic antibodies.