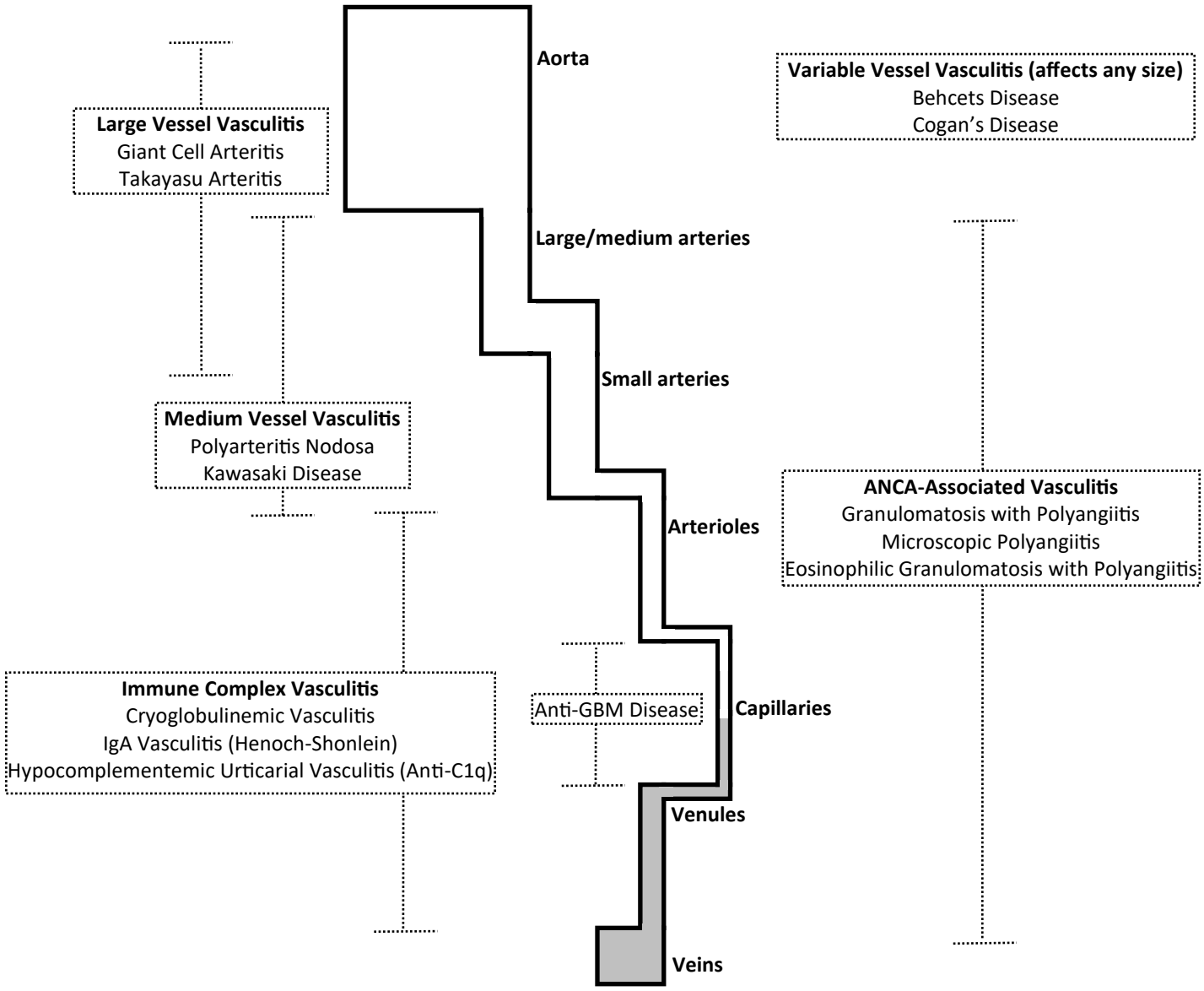
APPROACH TO VASCULITIS

BOTTOM LINE

Vasculitis is a heterogenous group of diseases classified by blood vessel size. Large vessel vasculitis symptoms include claudication, bruits, aneurysms, asymmetrical blood pressures/pulses. Medium vessel includes skin nodules/ulcers, livedo, bowel infarction, and digital necrosis. Small vessel includes petechiae/pupura, glomerulonephritis, alveolar hemorrhage, and mononeuritis multiplex. Specific vasculitis diagnoses affects a predictable size of blood vessel; each blood vessel size presents with predictable signs and symptoms of vasculitis. Thus, a careful history and exam can tell us which blood vessel size is affected, which vasculitis diagnoses may be responsible, and what confirmatory tests to order. Treatment is generally commensurate with degree of organ dysfunction.

CLASSIFICATION



Vessel size	Example vessels	Possible Symptoms/Signs
Large	Aorta and major branches (ex: carotid, subclavian, iliac)	Limb claudication, bruits, asymmetric blood pressures, pulse deficit, aortic aneurysm, cranial symptoms (temporal headache, jaw claudication, blindness)
Medium	Main visceral arteries/veins and initial branches (ex: renal, mesentery, deep skin, nerves)	Skin nodules and ulcers, livedo reticularis, digital necrosis, mononeuritis multiplex, renovascular hypertension, bowel infarction
Small	Intraparenchymal vessels (ex: glomeruli, alveoli, superficial skin, nerves)	Palpable purpura, GN, alveolar hemorrhage, scleritis, mononeuritis multiplex, urticaria >24h

DEFINITIONS (Chapel Hill Consensus Conference 2012)	
LARGE VESSEL VASCULITIS	
Takayasu arteritis (TAK)	Arteritis, often granulomatous, of aorta/major branches. Usually patients <50 years.
Giant cell arteritis (GCA)	Arteritis, often granulomatous, of aorta and/major branches—especially cranial (ex temporal), carotid, vertebral arteries. Patient >50 years (mean age ~70), PMR association.
MEDIUM VESSEL VASCULITIS	
Polyarteritis nodosa (PAN)	Necrotizing arteritis of medium/small arteries without glomerulonephritis; ANCA negative.
Kawasaki disease (KD)	Arteritis of medium and small arteries with the mucocutaneous lymph node syndrome. Coronary arteries, aorta and large arteries may be involved. Paediatric disease.
SMALL VESSEL VASCULITIS	
ANCA-associated vasculitis (AAV) Necrotizing pauci-immune (few/no immune deposits) vasculitis of small vessels with ANCA association in many.	
Microscopic polyangiitis (MPA)	Necrotizing pauci-immune small-vessel vasculitis; medium arteries may be involved. Necrotizing glomerulonephritis and pulmonary capillaritis common. No granulomas
Granulomatosis with Polyangiitis (GPA)	Necrotizing pauci-immune small/medium-vessel vasculitis usually involving upper/lower respiratory tract. Necrotizing glomerulonephritis is common.
Eosinophilic granulomatosis with polyangiitis (EGPA)	Necrotizing pauci-immune small/medium-vessel vasculitis with eosinophil-rich inflammation. Associated with asthma and other eosinophilia.
Immune complex vasculitis Vasculitis deposition of immunoglobulin and/or complement components on vessel wall, usually affecting small vessels. Glomerulonephritis is common.	
Anti-glomerular basement membrane (anti-GBM)	Vasculitis of glomerular capillaries or pulmonary capillaries (or both). Anti-GBM antibodies. Often presents with pulmonary hemorrhage/glomerulonephritis syndrome.
Cryoglobulinemic vasculitis (CV)	Vasculitis of small vessels with deposition of cryoglobulin; associated with serum cryoglobulins. Often presents with skin, glomeruli, and peripheral nerves involvement.
IgA vasculitis (Henoch-Schönlein) (IgAV)	Vasculitis of small vessels with IgA1-dominant immune deposits. Often presents with skin, GI, joint symptoms; can get glomerulonephritis.
Hypocomplementemic urticarial vasculitis (HUV, anti-C1q vasculitis)	Vasculitis/urticaria syndrome affecting small vessels; associated and hypocomplementemia and anti-C1q antibodies. Often presents with glomerulonephritis, arthritis, COPD, and ocular inflammation.
VARIABLE VESSEL VASCULITIS Vasculitis with no predominant type of vessel involved that can affect vessels of any size (small, medium, and large) and type (arteries, veins, and capillaries).	
Behçet's disease	Vasculitis affecting arteries or veins characterized by recurrent oral/genital ulcers. Often accompanied with skin, eye, joint, GI, and/or CNS inflammatory lesions. Small-vessel vasculitis, thromboangiitis, thrombosis, arteritis, and arterial aneurysms may occur.
Cogan's syndrome	Vasculitis characterized by eye and ear inflammation: keratitis, uveitis, episcleritis, sensorineural hearing loss and vestibular dysfunction. Other manifestations include arteritis, aortitis, aortic aneurysms, and aortic and mitral valvulitis.
SINGLE-ORGAN VASCULITIS Vasculitis of any size in a single organ (ex: isolated skin vasculitis, isolated renal vasculitis, primary CNS vasculitis).	
VASCULITIS ASSOCIATED WITH SYSTEMIC DISEASE Vasculitis associated or secondary to a systemic disease (ex: rheumatoid vasculitis, lupus vasculitis, malignancy, IBD).	
VASCULITIS ASSOCIATED WITH PROBABLE ETIOLOGY Vasculitis associated with a probable specific etiology (ex: drug-induced, hepatitis B-associated vasculitis).	

VASCULITS MIMICS	
Large arteries	• **Atherosclerosis** • Fibromuscular dysplasia • Congenital coarctation of aorta, middle aortic syndrome • Genetic diseases (Marfan syndrome, Ehlers-Danlos, Loeys–Dietz syndrome) • Infectious, acute: mycotic aneurysms associated with sepsis or endocarditis • Infectious, chronic: Syphilis, TB, HIV, leprosy • Isolated aortitis • IgG4 disease • Erdheim-Chester's disease
Medium arteries	• **Atherosclerosis** • Fibromuscular dysplasia • Genetic diseases (Marfan syndrome, Ehlers-Danlos) • Thromboembolic disease • Cholesterol emboli syndrome • Calciphylaxis • Segmental arterial mediolysis (SAM)
Cerebral arteries:	• RCVS • PRES • Cerebral amyloid angiopathy • Moyamoya disease • CADASIL • ADA2 deficiency/DADA2, Sneddon syndrome • CNS lymphoma
Small arteries	• Infectious endocarditis or other cardiogenic emboli, Atrial myxoma • Cholesterol microemboli syndrome • Thromboembolic disease • Anti-phospholipid antibody syndrome • Sepsis other infection • Cocaine • TTP • Calciphylaxis • Intravascular lymphoma

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