

Table 1 – Highlighted Conditions With Arteritis or Periarteritis of the Coronaries

Condition	Size Artery Involved	Estimated Annual Incidence	Common Clinical Characteristics	Common Laboratory Abnormalities	Frequency Coronary Involvement	Suggestive Coronary Angiographic Features	Suggestive Extra-Coronary Angiographic Features	Treatment
Takayasu's arteritis	Large (++) Medium (+)	1-2 per million	Onset < 40 yrs Limb claudication Arterial bruit Asymmetric pulse/BP	↑ ESR, CRP	10-30%	Ostial/proximal stenosis Skip lesions	Thickening ± narrowing/occlusion of large arteries (aorta or primary branches)	GC MTX TNF-inhibitor
Giant cell arteritis	Large (++) Medium (+)	10-30 / 100,000*	Onset ≥ 50 yrs Cranial symptoms (Headache, jaw claudication, double vision, vision loss)	↑ ESR, CRP	Rare	Tapered smooth narrowing Skip lesions	If large vessels involved: similar to Takayasu but aorta and subclavians > carotid, other	GC TCZ
Polyarteritis nodosa	Medium (++) Small (+)	4-10 per million	Skin nodules, livedo Abdominal pain Testicular pain (men) Mononeuritis multiplex	Hepatitis serologies (-) ANCA	10-50%	Aneurysm or alternating aneurysm/narrowing (beaded pattern)	-Visceral infarcts -Aneurysms (micro or saccular) -Alternating aneurysm/narrowing (beaded pattern)	GC CYC MTX
ANCA vasculitis	Medium (+) Small (++)	1-3 / 100,000	Recurrent sinusitis Pulmonary nodules Hemoptysis Glomerulonephritis Leukocytoclastic vasculitis	(+) p-ANCA/MPO or (+) c-ANCA/PR3 (+) Hematuria ↑ Creatinine	Rare	Non-specific	N/A	<u>Induction:</u> GC + (RTX or CYC) <u>Maintenance</u> RTX or AZA, MTX
Behcet's disease	Variable	0.2-80 / 100,000*	Oral and genital sores Ulceration GI tract Ocular inflammation Venous/arterial thrombosis	N/A	0.5-2%	Non-specific	Venous and/or arterial thrombosis Pseudoaneurysm medium/large arteries	Colchicine GC CYC, MTX TNF inhibitor
IgG4-related disease	Variable	0.3-1.0 / 100,000	Lymphadenopathy Salivary/Lacrimal swelling Orbital pseudotumor Retroperitoneal fibrosis Autoimmune pancreatitis	↑ IgG4 (±)	1-3%	Aneurysm Periarteritis	Periaortitis	GC RTX
Erdheim-Chester disease	Variable	Rare, unknown	Bone pain (osteosclerosis) Right atrial pseudotumor Exophthalmos Perirenal fibrosis Xanthelasma	BRAF V600E mutation on histology	25-55%	Periarteritis (RCA > LM > other)	Periaortitis "Coated Aorta"	Interferon Cladribine BRAF-inhibitor

ANCA, anti-neutrophil cytoplasmic antibody; AZA, azathioprine; BP, blood pressure; CRP, C-reactive protein; CYC, cyclophosphamide; ESR, erythrocyte sedimentation rate; GC, glucocorticoids; IgG4, immunoglobulin G4; LM, left main; MTX, methotrexate; RCA, right coronary artery; RTX, rituximab; TCZ, tocilizumab; TNF, tumor necrosis factor

*GCA annual incidence estimate is among patients aged ≥50 years of age; †Behcet's disease incidence highest in Middle East and Turkey