

Table 1. Comparison of Demographic Features, Presentation, and Histopathology in the Most Common Causes of Aortitis

Etiology	Demographics	Location of Disease	Distinct Associated Symptoms	Findings on Histopathology ²³
Takayasu's arteritis ^{8,9,26,31}	Overall: Asia > Northern European descent Type I, II most common in Japanese and Northern European descent Type III, IV, V most common in South Asian and African descent Women-to-men ratio = 8.5:1 Age <40 y	- Type I—branches of aortic arch - Type IIa—ascending aorta, and aortic arch and its branches - Type IIb—ascending aorta, and aortic arch and its branches, and thoracic descending aorta - Type III—thoracic descending, abdominal or renal arteries - Type IV—abdominal aorta or renal arteries - Type V—combined features of IIb and IV	Aortic valve thickening with aortic insufficiency, pulmonary artery aneurysm, or stenosis	Clusters of epithelioid macrophages with or without giant cells. Often granulomas present. Outer media and adventitia > inner media.
Giant cell arteritis ^{12,13,32}	Overall: Northern European > Southern European Caucasian > Asian, Black Women-to-men ratio = 2-3:1 Age >50 y	- Thoracic > abdominal aorta - Stenoses mostly limited to the aortic arch and branch vessels - Temporal artery (temporal arteritis) - Polymyalgia rheumatica	Headaches, jaw claudication, proximal joint stiffness and pain	Clusters of epithelioid macrophages with or without giant cells. Rarely with compact, well-formed granulomas present. Inner media > outer media and adventitia
IgG4-related aortitis ^{15,19,33}	Men > women Mean age diagnosis 51-58 years	- Abdominal > thoracic aorta > other - Type 1—ascending aorta, arch or descending thoracic aorta - Type 2a—abdominal aorta - Type 2b—abdominal aorta and iliac arteries - Type 2c—iliac arteries only - Type 3—thoracic and abdominal aorta involvement	Hydronephrosis or ureteral obstruction	Lymphocytes and plasma cells with a granulomatous component; obliterative adventitial phlebitis; fibrosis (may be storiform). IgG4 ⁺ plasma cells >50 per 400× HPF and IgG4/IgG ratio >50%. Lymphoplasmacytic aortitis or limited to adventitia in periaortitis.
Isolated aortitis ^{14,15}	Men > women Mean age of diagnosis 62 y	Aortic arch, thoracic or abdominal aorta	No branch vessel involvement	Most often granulomatous inflammation that is indistinguishable from giant cell arteritis. May be purely lymphoplasmacytic and, on routine slides, indistinguishable from IgG4-related aortitis.

HPF = high-power field; Ig = immunoglobulin.

Kadian-Dodov D, Seo P, Robson PMet al. Inflammatory Diseases of the Aorta: JACC Focus Seminar, Part 2. *JACC*. 2022;80:832-844.

Table 2. ACC/AHA 2022 GL on Aortic Disease. Inflammatory Aortitis: Diagnosis and Treatment of Takayasu Arteritis and Giant Cell Arteritis (GCA).

Diagnosis

In patients with large vessel vasculitis (LVV), prompt evaluation of the entire aorta and branch vessels with MRI or CT, with or without 18F-FDG positron emission tomography (FDG-PET). I C-LD

Treatment

In patients with active GCA or Takayasu arteritis, initial medical therapy with high-dose glucocorticoids. I B-NR

In patients with GCA who have evidence of active aortitis, tocilizumab as adjunctive therapy to glucocorticoids, with methotrexate as an alternative. I B-R

In all patients with Takayasu arteritis, nonbiological disease-modifying anti-rheumatic drugs (DMARD) in combination with glucocorticoids. I C-LD

In patients with active GCA or Takayasu arteritis, treatment efficacy periodically assessed by monitoring inflammatory serum markers (C-reactive protein and erythrocyte sedimentation rate), imaging with CT, MRI, or FDG-PET, and clinical symptoms. I C-LD

In patients with GCA or Takayasu arteritis who are in remission, elective endovascular or open surgical intervention to treat aortic and branch vessel complications. IIa C-LD

In patients with GCA or Takayasu arteritis and aortic involvement who are in remission, Annual surveillance imaging with CT, MRI, or FDG-PET. IIa C-EO

Diagnostic Criteria for Inflammatory Aortitis

Names	Criteria Used in Diagnosis	When Is Diagnosis Established?
Takayasu arteritis	Age of onset <40 y	≥3 criteria are present (sensitivity 90.5%; specificity 97.8%)
	Intermittent claudication	
	Diminished brachial artery pulse	
	Subclavian artery or aortic bruit	
	Systolic BP variation of >10 mm Hg between arms	
	Aortographic evidence of aorta or aortic branch stenosis	
Giant cell arteritis	Age >50 y	≥3 criteria are present (sensitivity >90%; specificity >90%)
	Recent-onset localized headache	
	Temporal artery tenderness or pulse attenuation	
	Elevated erythrocyte sedimentation rate >50 mm/h	
	Arterial biopsy shows necrotizing vasculitis	

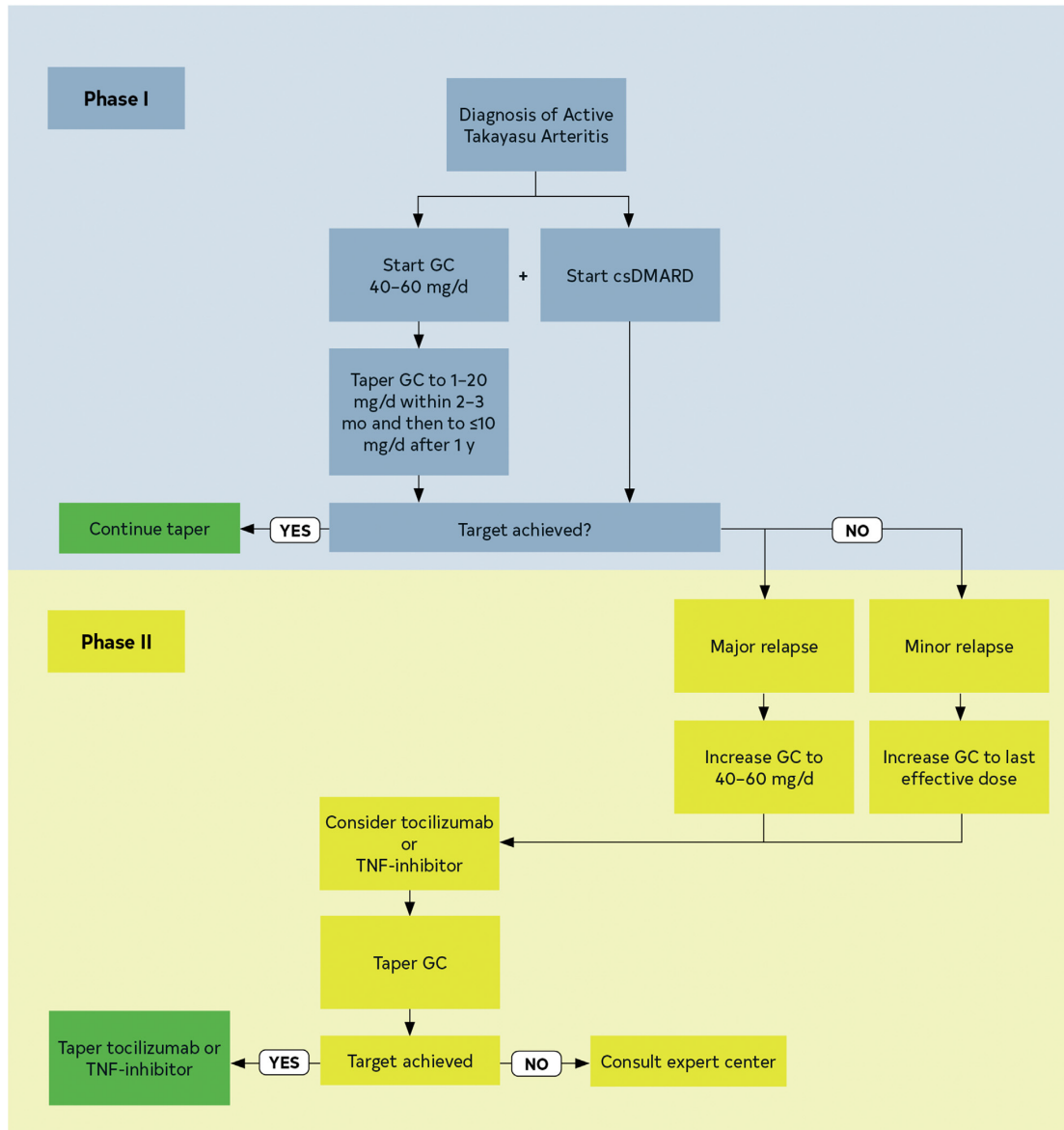


Figure 1. ACC/AHA 2022 GL on Aortic Disease. The 2018 European Alliance of Associations for Rheumatology (EULAR; formerly European League Against Rheumatism) Recommended Algorithms for the Pharmacological Treatment of Takayasu Arteritis.

csDMARD indicates conventional synthetic disease modifying antirheumatic drug; GC, glucocorticoids; and TNF, tumor necrosis factor.

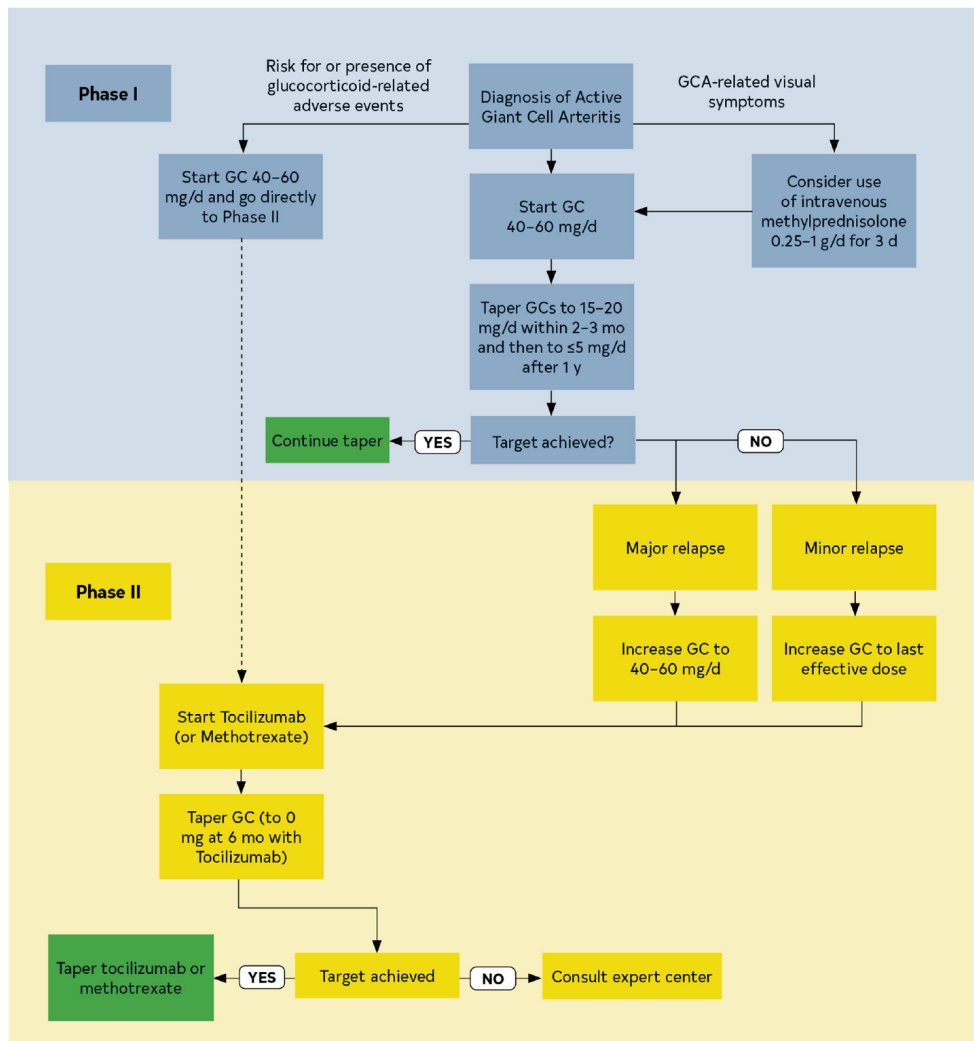


Figure 2. ACC/AHA 2022 GL on Aortic Disease. The 2018 European Alliance of Associations for Rheumatology (EULAR; formerly European League Against Rheumatism) Recommended Algorithms for the Pharmacological Treatment of Giant Cell Arteritis

GC indicates glucocorticoids; GCA, giant cell arteritis; and TNF, tumor necrosis factor.

Table 3. ACC/AHA 2022 GL on Aortic Disease.**Diagnosis and Management of Infection of the Native Aorta**

Open surgical repair in patients with infectious aortitis and associated aneurysms or dissection of the thoracic or abdominal aorta.	I C-EO
In select patients, treatment with endovascular repair.	IIb C-LD
In patients with infectious aortitis complicated by rupture, either open or endovascular repair, based on the patient's status at presentation and institutional expertise.	IIa C-EO
In patients with infectious aortitis, intravenous antimicrobial therapy of at least 6 weeks' duration, with lifelong suppressive therapy in select cases not amenable to interventional repair or who have recurrent infection.	IIb C-EO

Management of Aortic Mycotic Aneurysm: Comparison of Resection and Extra-Anatomic Reconstruction, In Situ Reconstruction, or Endovascular Device Repair

Procedure	Potential Indications*	Advantages	Disadvantages
Extra-anatomic reconstruction	Infrarenal location with gross purulence, psoas or retroperitoneal abscess, vertebral osteomyelitis, inadequate response to antibiotic therapy, selected aortoenteric fistulae	Avoids placement of foreign body in infected area	Not technically feasible for thoracic, suprarenal, or visceral location or for emergency use Long operating time Long-term patency rates low Stump blowout Limb ischemia, amputation Reinfection rate higher than for in situ reconstruction Ischemic colitis
In situ reconstruction	Thoracic, suprarenal, infrarenal, or visceral location Selected aortoenteric fistulae	More versatile than extra-anatomic: fewer long-term complications, higher patency rates, lower recurrent infection rate, shorter operating time Polyester grafts† available for emergency surgery	Theoretical risk of infection because of interposition of foreign material in infected site
Endovascular device repair	Bridge procedure‡: hemodynamic instability, uncontrolled bleeding, rupture or impending rupture, selected patients with aortocentric fistulae, patients who are not fit for open surgery	Emergency stabilization Low early morbidity, mortality Less invasive No cross-clamping of aorta: spinal cord injury, reperfusion injury	Persistent infections and device infections Higher long-term morbidity, mortality with device retention Requires device explanation, reconstruction

*Potential indication; must be individualized for each patient.

†Polyester grafts, rifampin-soaked or silver-coated; less experience reported with cryopreserved arterial allografts or venous autografts.

‡Bridge procedure, used to stabilize patients until device explanation and arterial reconstruction.

Diagnosis and Management of Prosthetic Aortic Graft Infection**Diagnosis**

In patients with a prosthetic aortic graft, who have signs and symptoms or culture evidence of unexplained infection or have unexplained gastrointestinal bleeding, cross-sectional imaging to evaluate for an underlying aortic graft infection.	IIa B-NR
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Treatment

In patients with an infected prosthetic aortic graft who are hemodynamically	IIa B-NR
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stable and have appropriate anatomy, perform open surgery with either in situ reconstruction or extra-anatomic bypass.

In patients with an infected prosthetic aortic graft who are hemodynamically unstable, perform open surgery with either explant or in situ reconstruction. IIa B-NR

In patients with an infected prosthetic aortic graft, endovascular therapy either as bridge therapy in those with hemodynamic instability or as long-term therapy in those who are unsuitable candidates for open surgery. IIa C-LD

Late Management

In patients who have undergone treatment of an acute prosthetic aortic graft infection, targeted intravenous antimicrobial therapy of at least 6 weeks' duration, with prolonged suppressive oral therapy in select cases, plus a consultation and follow-up with an infectious disease specialist. I C-LD

In patients with an infected prosthetic aortic graft and either an extensive perigraft abscess or an infection caused by methicillin-resistant *S. aureus*, *Pseudomonas aeruginosa*, or a multidrug-resistant microorganism, or who have undergone in situ reconstruction, lifelong suppressive oral antimicrobial therapy after the initial course of therapy. IIb C-LD