The Journal of Rheumatology 2021;48:1583–8
doi:10.3899/jrheum.201274
First Release July 1 2021

The Journal of Rheumatology

Spectrum and Outcome of Noninfectious Aortitis

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ABSTRACT. ***Objective.*** To assess the spectrum and long-term outcome of patients with noninfectious aortitis.

Methods. We performed a retrospective multicenter study of 353 patients (median age at diagnosis was 62 [IQR 46–71] yrs and 242 [68.6%] patients were women) with noninfectious aortitis. Factors associated with vascular complications were assessed in multivariate analysis.

Results. We included 136 patients with giant cell arteritis (GCA), 96 with Takayasu arteritis (TA), 73 with clinically isolated aortitis (CIA), and 48 with aortitis secondary to inflammatory diseases (including Behçet disease, relapsing polychondritis, IgG4-related disease, Cogan syndrome, ankylosing spondylitis). After a median follow-up of 52 months, vascular complications were observed in 32.3%, revascularizations in 30% of patients, and death in 7.6%. The 5-year cumulative incidence of vascular complications was 58% (95% CI 41–71), 20% (95% CI 13–29), and 19% (95% CI 11–28) in CIA, GCA, and TA, respectively. In multivariate analysis, male sex (HR 2.10, 95% CI 1.45–3.05, $P < 0.0001$) and CIA (HR 1.76, 95% CI 1.11–2.81, $P = 0.02$) were independently associated with vascular complications.

Conclusion. Noninfectious aortitis accounts for significant morbidity and mortality. CIA seems to carry the highest rate of vascular complications.

Key Indexing Terms: aortitis, noninfectious aortitis, large vessel vasculitides

Noninfectious aortitis is presumed to be an autoimmune disease. The most common causes of aortitis are the large-vessel vasculitides: giant cell arteritis (GCA) and Takayasu arteritis (TA).[1] Although it may occur as part of another systemic autoimmune/inflammatory disease (including rheumatoid arthritis, spondyloarthritis, Behçet disease [BD], Cogan syndrome, systemic lupus erythematosus, Sjögren syndrome, granulomatosis with polyangiitis, sarcoidosis, or IgG4-related disease), it may also appear as isolated, as a topographically limited lesion. The term *clinically isolated aortitis* (CIA) was included in the Chapel Hill classification under the category of single organ vasculitis.[1]

Aortitis is diagnosed by imaging tools of the entire aorta including computed tomography (CT) angiography and magnetic resonance angiography (MRA). Positron emission tomography (PET) scanning has emerged for targeted imaging of vascular inflammation and may be particularly useful when combined with traditional cross-sectional imaging modalities.[1,2,3] Alternatively, aortitis may be found on histopathological examination after surgery for aorta aneurysm or dissection.

Aortitis is related to significant morbidity and mortality through the development of aortic aneurysm, aortic wall rupture, aortic acute dissection, and/or thrombotic luminal obstruction.[3] Because of the wide variation in the course of aortitis, predicting outcome is challenging. The objective of our study was to identify prognosis factors in complications of noninfectious aortitis to define high-risk patients, by using a multivariate model.

METHODS

Patients. We conducted a retrospective multicenter study in referral centers from the French Study Group for Large Vessel Vasculitis (GEFA) between 2000 and 2016. We identified 353 patients with noninfectious aortitis. Aortitis was diagnosed either through histopathology, when active inflammatory infiltrates that involve the intima and/or media was demonstrated, or radiographically, as circumferential aortic wall thickening on magnetic resonance imaging or CT scans with or without corresponding increased fluodeoxyglucose (FDG) uptake on PET scans. We identified 136 patients with GCA fulfilling the 1990 American College of Rheumatology (ACR) diagnostic criteria for GCA,[4] 96 patients with TA fulfilling the TA ACR[5]

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DS has received consulting and lecturing fees from Medimmune, AbbVie, Bristol Myers Squibb, Amgen, Celgene, Sanofi-Genzyme, Roche-Chugai, Servier, Gilead, AstraZeneca, and GlaxoSmithKline. PC has received consulting and lecturing fees from AbbVie, AstraZeneca, Bristol Myers Squibb, Gilead, GlaxoSmithKline, Janssen, MSD, Roche, Servier, and Vifor. The remaining authors declare no conflicts of interest relevant to this article.

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Accepted for publication April 12, 2021.

and/or Ishikawa criteria modified by Sharma *et al*,[6] and 73 patients with CIA defined by histological inflammation and/or imaging in any part of the aorta in the absence of an underlying defined inflammatory condition as defined in revised Chapel Hill vasculitis nomenclature.[1] GCA, TA, and CIA were defined at the time of diagnosis of aortitis. For patients with CIA, the absence of an underlying defined inflammatory condition was checked at the end of follow-up. All patients underwent vascular imaging during follow-up. The nature and frequency of imaging (CT angiography, MRA, and/or FDG-PET) was left at the discretion of the physician in charge of the patients. However, since patients were recruited from tertiary centers, imaging monitoring was carried out at least once a year.

The study was approved by the local ethics committee (Paris VI Ethics Committee, ethics approval number: 1867484). We reviewed the electronic medical records of all patients that included age at diagnosis of aortitis, etiology, race, sex, topography of arterial lesion (ascending aorta, arch of aorta, descending aorta, abdominal aorta), imaging features (CT scan, MRI, FDG-PET scan), histological data, treatments (corticosteroids, immunosuppressive treatments), vascular complications (dissection or aneurysms), vascular revascularization including open or endovascular aortic procedures (histopathology results of additional vascular tissue obtained were recorded), and death.

Definitions of study endpoints. Vascular complications were defined as the occurrence of one of these events: aneurysm and/or dissection. Aneurysm was defined as an outer aortic diameter > 3 cm or > 50% of the normal diameter of a healthy individual of the same sex and age. Revascularizations were defined as open or endovascular aortic procedures, endarterectomies, branch vessel bypasses or repairs, and angioplasties ± stenting.

Statistical analysis. Continuous variables are presented as median (IQR), and categorical variables are presented as number (%). The *P* values reported in Table 1 correspond to statistical 2-sided tests of null hypotheses defined as the distribution of the variables being identical across all 4 groups. The corresponding alternative hypotheses are that the distributions differ according to the group, overall. In that sense, the *P* values apply to the grouping variable as a whole; *P* values < 5% were considered to indicate that the variable distribution (e.g., sex, age) was not independent from the group (GCA, TA, CIA, or other). Vascular event–free survival was defined as the time from the date of aortitis diagnosis to the date of the first vascular complication (aneurysm or dissection). Event-free survival was defined as the time from the date of aortitis diagnosis to the date of the first vascular complication (aneurysm or dissection), first revascularization, or death (of any cause), whichever occurred first. Survival functions were estimated using the Kaplan-Meier method. Cumulative incidence of vascular events was computed, with death without vascular event as a competing event. Factors associated with time to event outcomes were evaluated using Cox models, with estimation of HRs and their 95% CIs. The proportional hazards assumption was assessed with examination of Schoenfeld residuals and Grambsch and Therneau test. For each outcome, variables associated with the outcome at a 0.05 level in univariate analyses were candidates for multivariate analysis. The selection of multivariate-adjusted models was based on the minimization of Akaike criterion, with a stepwise selection procedure. All tests were 2-sided; *P* values < 0.05 indicated significant associations. Analyses were performed using R version 3.5.3. (R Foundation for Statistical Computing).

RESULTS

Characteristics of the study population. The main clinical and demographic characteristics of the 353 patients with noninfectious aortitis at diagnosis are presented in Table 1. The median age at diagnosis was 62 (IQR 46–71) years and 242 (68.6%) patients were women. Patients were predominantly White (76.2%). Eighty (22.7%) patients had a histological diagnosis of aortitis. The topography of aortitis vessel lesions included descending thoracic aorta involvement in 72.2%, aortic arch involvement in 64.3%, and ascending aortic involvement in 59.8% of cases. Aortitis secondary to autoimmune/inflammatory

Table 1. Characteristics of patients with noninfectious aortitis.

	Total, n = 353	GCA, n = 136	TA, n = 96	CIA, n = 73	Other, n = 48	*P*
Female sex	242 (68.6)	102 (75.0)	75 (78.1)	44 (60.3)	21 (43.8)	< 0.0001
Age at diagnosis, yrs, median (IQR)	62 (46–71)	70 (64–76)	39 (25–49)	64 (54–71)	58 (37–64)	< 0.0001
Race						< 0.0001
White	269 (76.2)	129 (94.9)	43 (44.8)	66 (90.4)	31 (64.6)	
Non-White	84 (23.8)	7 (5.1)	53 (55.2)	7 (9.6)	17 (35.4)	
Etiology						
Behçet disease	16 (4.5)	0 (0.0)	0 (0.0)	0 (0.0)	16 (33.3)	
Relapsing polychondritis	10 (2.8)	0 (0.0)	0 (0.0)	0 (0.0)	10 (20.8)	
IgG4-related disease	7 (2.0)	0 (0.0)	0 (0.0)	0 (0.0)	7 (14.6)	
Cogan syndrome	2 (0.6)	0 (0.0)	0 (0.0)	0 (0.0)	2 (4.2)	
Ankylosing spondylitis	2 (0.6)	0 (0.0)	0 (0.0)	0 (0.0)	2 (4.2)	
Sarcoidosis	1 (0.3)	0 (0.0)	0 (0.0)	0 (0.0)	1 (2.1)	
Others	10 (2.8)	0 (0.0)	0 (0.0)	0 (0.0)	10 (20.8)	
Topography of arterial lesions						
Descending aorta	255 (72.2)	109 (80.1)	70 (72.9)	53 (72.6)	23 (47.9)	0.0006
Abdominal aorta	230 (65.2)	91 (66.9)	63 (65.6)	46 (63.0)	30 (62.5)	0.92
Arch of aorta	227 (64.3)	91 (66.9)	67 (69.8)	50 (68.5)	19 (39.6)	0.003
Ascending aorta	211 (59.8)	90 (66.2)	53 (55.2)	47 (64.4)	21 (43.8)	0.03
Treatment						
Corticosteroids[a]	330 (93.5)	135 (99.3)	92 (95.8)	65 (89.0)	38 (79.2)	
Immunosuppressants[a]	153 (43.3)	27 (19.9)	71 (74.0)	18 (24.7)	37 (77.1)	
Vascular procedure	106 (30.0)	29 (21.3)	37 (38.5)	24 (32.9)	16 (33.3)	

Data are expressed as n (%) unless otherwise indicated. [a] Baseline treatment initiated within 1 month of diagnosis. CIA: clinically isolated aortitis; GCA: giant cell arteritis; TA: Takayasu arteritis.

diseases included BD (n = 16), relapsing polychondritis (n = 10), IgG4-related disease (n=7), Cogan syndrome (n = 2), ankylosing spondylitis (n = 2), sarcoidosis (n = 1), and others (histiocytosis [n = 5], leukemia [n = 2], granulocyte colony stimulating factor–associated aortitis [n = 1], myelodysplasia [n = 1], primary biliary cholangitis [n = 1]). Most patients received corticosteroids (93.5%) and 153 (43.3%) patients received immunosuppressive therapy. The median (IQR) follow-up was 52 (25–92), 71 (32–137), and 29 (15–57) months in GCA, TA, and CIA, respectively. At the time of diagnosis, patients in the CIA group did not have more aneurysms than the others.

Factors associated with vascular complications. Vascular complications were observed in 114 (32.3%) patients. One hundred seven (30.3%) patients had aneurysms, 4 (1.1%) patients had false aneurysms or arterial ulcerations, and 18 (5.1%) had dissections (Table 2). The 3-, 5-, and 10-year cumulative incidence of vascular complications was 32% (95% CI 21–44), 58% (95% CI 41–71), and 71% (95% CI 50–85) in CIA; 15% (95% CI 9–22), 20% (95% CI 13–29), and 45% (95% CI 30–60) in GCA; and 11% (95% CI 6–19), 19% (11–28), and 35% (95% CI 22–48) in TA, respectively (Figure 1). In univariate analysis, male sex ($P < 0.0001$), age > 50 years ($P = 0.002$), TA ($P = 0.009$), and CIA ($P = 0.004$) were associated with vascular complications, compared to GCA (Table 3). In multivariate analysis, male sex ($P < 0.0001$) and CIA ($P = 0.02$, vs GCA) were independently associated with higher risk of vascular complications (Table 3).

Factors associated with vascular procedures. Revascularization was performed in 106 (30%) patients. The 1-, 3-, 5-, 10-year revascularization-free survival rates were 91% (95% CI 88–94), 80% (95% CI 75–85), 69% (95% CI 64–75), and 53% (95% CI 45–61), respectively. In univariate analysis, male sex ($P = 0.002$) and CIA ($P = 0.02$, vs GCA) were associated with revascularization. In multivariate analysis, male sex ($P = 0.004$) was independently associated with revascularization, whereas CIA was associated with higher risk of revascularization compared to GCA ($P = 0.06$; Figure 2).

DISCUSSION

In this large study, we assessed the spectrum and long-term outcome of noninfectious aortitis. We analyzed prognostic factors associated with the occurrence of vascular complications and revascularizations. The most striking conclusions drawn from this study are these: (1) 50% of patients with noninfectious aortitis will experience a vascular complication within 10 years following the diagnosis; and (2) the incidence of vascular complications after a 5-year follow-up is 2.9-times higher in CIA compared to GCA and TA. Although multiple series have identified idiopathic aortitis as an incidental histopathological finding after aortic surgery,[7] clinical outcomes in these patients have been difficult to compare due to considerable heterogeneity in definitions of clinical phenotypes, approaches to treatment, monitoring, and duration of follow-up.

Corticosteroids were prescribed to the same extent in patients with CIA, GCA, and TA. Although patients with CIA and GCA received less immunosuppressive therapy compared to those with TA, neither steroids nor immunosuppressants were independently associated with vascular events or vascular procedure in multivariate analysis.

In a small prospective study of 16 patients with aortitis, vascular events were 2 times higher in CIA compared to patients with secondary aortitis.[8] The rate of aortic events at 34 months was also markedly higher (55% vs 27%) in CIA compared to

Table 2. Outcomes of patients with noninfectious aortitis.

	Total, n = 353	GCA, n = 136	TA, n = 96	CIA, n = 73	Other, n = 48
Vascular events					
Aneurysms	107 (30.3)	29 (21.3)	24 (25.0)	31 (42.5)	23 (47.9)
False aneurysms or arterial ulceration	4 (1.1)	1 (0.7)	2 (2.1)	0 (0.0)	1 (2.1)
Dissection	18 (5.1)	7 (5.1)	2 (2.1)	7 (9.6)	2 (4.2)
Death	27 (7.6)	20 (14.7)	0 (0.0)	5 (6.8)	2 (4.2)
Cause of death					
Unknown	6 (22.2)	4 (20.0)	–	1 (20.0)	1 (50.0)
Aortic aneurysm	4 (14.8)	2 (10.0)	–	1 (20.0)	1 (50.0)
Stroke	2 (7.4)	2 (10.0)	–	0 (0.0)	0 (0.0)
Septicemia	2 (7.4)	2 (10.0)	–	0 (0.0)	0 (0.0)
Aortic dissection	2 (7.4)	2 (10.0)	–	0 (0.0)	0 (0.0)
Myocardial infarction	2 (7.4)	1 (5.0)	–	1 (20.0)	0 (0.0)
Mesenteric ischemia	2 (7.4)	0 (0.0)	–	2 (40.0)	0 (0.0)
Heart failure	2 (7.4)	2 (10.0)	–	0 (0.0)	0 (0.0)
Cardiogenic shock	1 (3.7)	1 (5.0)	–	0 (0.0)	0 (0.0)
Pulmonary embolism	1 (3.7)	1 (5.0)	–	0 (0.0)	0 (0.0)
Cerebral hemorrhage	1 (3.7)	1 (5.0)	–	0 (0.0)	0 (0.0)
Peritonitis	1 (3.7)	1 (5.0)	–	0 (0.0)	0 (0.0)
Pneumonitis	1 (3.7)	1 (5.0)	–	0 (0.0)	0 (0.0)

Data are expressed as n (%) unless otherwise indicated. CIA: clinically isolated aortitis; GCA: giant cell arteritis; TA: Takayasu arteritis.

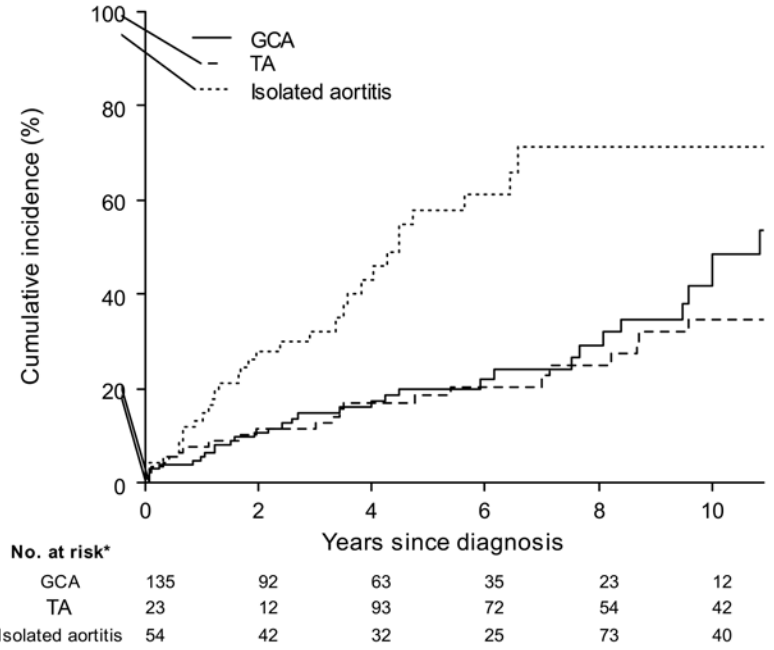

Figure 1. Cumulative incidence of vascular complications by cause of aortitis. GCA: giant cell arteritis; TA: Takayasu arteritis.

patients with GCA in a cohort of 117 patients.[9] Clifford, *et al* highlighted the occurrence of 45% new vascular lesions and 40% vascular revascularization during follow-up in patients with CIA.[10]

Currently, the mainstay therapy for large-vessel vasculitis is glucocorticoids (GCs).[11] However, the efficacy of GCs in CIA remains unclear. In the present study, 89.0% and 24.7% of patients with CIA received corticosteroids and immunosuppressive treatment, respectively. In a previous cohort of patients with CIA, 6/25 (24%) of untreated patients compared to 0/11 (0%) of those receiving steroids developed aneurysms.[12] However, other studies did not find a benefit of using steroids in CIA.[11,13,14]

In the present study, the 5-year revascularization-free survival was 41% in CIA compared to 81% and 74% in TA and GCA, respectively. This frequency is comparable to that reported previously,[10,11,12,13] and highlights the need for special monitoring and for continued surveillance in patients with CIA. In a French retrospective multicenter study comparing 73 patients with GCA and 44 with CIA, aortic surgery was more frequently performed in the CIA group (63.4% vs 13.7%).[9] A retrospective study from the Cleveland Clinic identified 196 patients with histopathology-proven aortitis among patients undergoing thoracic aortic surgery.[12] Patients met criteria for CIA (129 [66%]), GCA (42 [21%]), TA (14 [7%]) or other systemic inflammatory diseases (11 [6%]). During a mean follow-up of 56.2 months, 40% of patients with CIA underwent additional vascular surgery, although this was not statistically different with other subgroups.[12]

Our study brings to light important new information regarding the long-term outcome and prognosis of aortitis. CIA is an uncommon disorder characterized by giant cells or lymphoplasmacytic inflammation of the aorta. CIA appears as a multifocal and progressive aortic disease. The high prevalence of vascular complications reported in our study is in line with previous series.[12,13] It is important to underline that in this paper, most patients were diagnosed radiographically, whereas in the previous series aortitis was diagnosed histologically after surgical resection was performed.[7,8,10–14]

We acknowledge some limitations to our study. Our analysis was performed as a retrospective study. We were unable to collect complete longitudinal data of patients who were seen only on an intermittent consultation basis. Prospective enrollment and data collection from the time of diagnosis would have been ideal but difficult to achieve with such rare diseases. However, all patients have been followed in reference centers in which clinical, imaging, and treatment strategy are homogeneous.

In conclusion, this large study shows that 50% of patients with noninfectious aortitis will experience a vascular complication within 10 years following initial diagnosis. We identified male sex and CIA as risk factors for vascular complications. CIA seems to carry a 3-fold higher rate of vascular complications compared to TA and GCA. Together, our study provides critical information on the prognosis of patients with noninfectious aortitis that may serve to necessitate more aggressive management and closer follow-up. It remains unclear whether patients with CIA would benefit from GCs or other immunomodulatory therapies such as tocilizumab. Prospective studies are warranted to evaluate the efficacy of immunosuppressive therapy in CIA.

Table 3. Predictive factors of vascular complications.

	N Event/N	HR (95% CI)	*P*	Adjusted HR (95% CI)	*P*
Total	125/349				
Sex					
Female	66/239	1		1	
Male	59/110	2.32 (1.63–3.31)	< 0.0001	2.10 (1.45–3.05)	< 0.0001
Age, yrs					
≤ 50	34/103	1		1	
> 50	91/246	1.91 (1.27–2.88)	0.002	1.40 (0.81–2.43)	0.23
Race					
White	96/266	1			
Non-White	29/83	0.75 (0.49–1.13)	0.17		
Etiology					
GCA	43/135	1		1	
TA	25/93	0.51 (0.30–0.84)	0.009	0.66 (0.34–1.28)	0.22
CIA	34/73	1.95 (1.24–3.08)	0.004	1.76 (1.11–2.81)	0.02
Other	23/48	1.24 (0.74–2.07)	0.41	1.11 (0.62–2.00)	0.72
Descending aorta					
No	49/98	1			
Yes	76/251	0.63 (0.44–0.90)	0.01		
Abdominal aorta					
No	55/122	1			
Yes	70/227	0.74 (0.52–1.05)	0.09		
Arch of aorta					
No	58/125	1			
Yes	67/224	0.76 (0.53–1.08)	0.13		
Ascending aorta					
No	45/140	1			
Yes	80/209	1.15 (0.80–1.66)	0.45		
Corticosteroids					
No	15/41	1			
Yes	79/256	1.39 (0.78–2.49)	0.26		
Immunosuppressant					
No	20/62	1			
Yes	15/44	1.22 (0.61–2.45)	0.57		

CIA: clinically isolated aortitis; GCA: giant cell arteritis; TA: Takayasu arteritis.

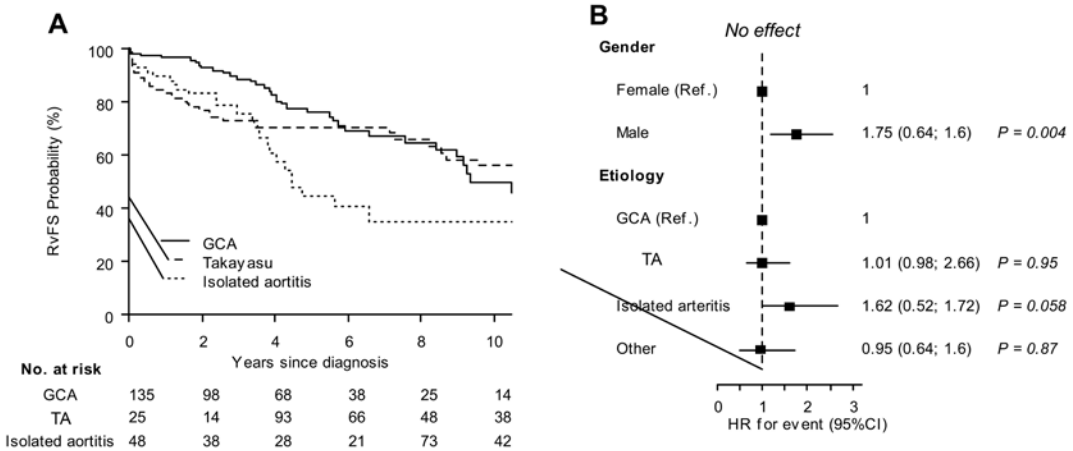

Figure 2. Revascularization-free survival (RvFS). (A) Kaplan-Meier estimates; and (B) multivariate model. GCA: giant cell arteritis; TA: Takayasu arteritis.

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