Survey: Assessing Canadian Practice Patterns Regarding Idiopathic Aortitis

SECTION 1: DEMOGRAPHICS

1. What is your age?

☐ < 35
☐ 35-45
☐ 46-55
☐ 56-65
☐ >65

2. Where is your practice based?

Province

City

3. What is your specialty?

☐ Trainee (resident in rheumatology)
☐ Adult Rheumatology
☐ Pediatric rheumatology
☐ Primary care
☐ Internal medicine
☐ Immunology
☐ Other (specify) _____

4. How many years have you practiced your specialty?

☐ Currently in residency or fellowship program
☐ 1 to 5 years
☐ 6 to 10 years
☐ 11 to 20 years
☐ 21 to 29 years
☐ Greater than 30 years

SECTION 1: DEMOGRAPHICS (continued)

5. How would you best describe your practice setting?

□ Solo community practice
□ Group community practice
□ Community practice affiliated with an academic centre
□ Clinical trials research facility associated with a community practice
□ Academic/teaching hospital
□ Other (please specify) _____

6. Are you a core or an associate member of a vasculitis research network? (CanVasc, VCRC, etc.)

□ Yes
□ No

7. How many patients with idiopathic or isolated aortitis have you seen over the whole course of your practice?

□ 0 (if selected, the survey ends at this point)
□ 1-3
□ 4-9
□ 10-19
□ 20 or more

SECTION 2: DEFINITIONS

The following section asks questions to assess how you define idiopathic aortitis and isolated aortitis in your practice.

When completing this section, please note: when we refer to a "**defined systemic inflammatory condition,**" we mean any systemic condition (as defined by the current nomenclature, classification or diagnostic criteria) known to be associated with aortitis, including: Behçet's disease, Cogan's syndrome, Giant cell arteritis (GCA), Granulomatosis with polyangiitis, Inflammatory bowel disease, Kawasaki disease, Polyarteritis nodosum, Polymyalgia Rheumatica, relapsing polychondritis, Rheumatoid arthritis, Sarcoidosis, Sjogren syndrome, spondyloarthropathies, Systemic lupus erythematosus, and Takayasu's arteritis (TAK).

8. Please select which of the attributes you consider to be part of the definition of idiopathic aortis:

Attribute	Required for definition of idiopathic aortitis	Irrelevant (ie you do not consider this attribute when deciding whether patient has idiopathic aortitis)
Have to exclude the presence of a defined systemic inflammatory condition (per existing diagnostic or classification criteria)	☐	☐
Have to exclude the presence of any of the clinical features suggesting a defined systemic inflammatory condition (eg – inflammatory arthritis, weight loss, inflammatory bowel disease)	☐	☐
Inflammatory markers should be normal	☐	☐
Have to exclude the presence of radiographic abnormalities in aortic branch vessels (eg – aneurysms, stenosis, thickening of vessel wall)	☐	☐
Other (please specify)	☐	☐
Other (please specify)	☐	☐

9. Do you make a distinction between idiopathic aortitis and isolated aortitis?

☐ No (automatically moved onto question 10)
☐ Yes (automatically moved onto question 9a)

SECTION 2: DEFINITIONS (continued)

When completing this section, please note: when we refer to a "**defined systemic inflammatory condition,**" we mean any systemic condition (as defined by the current nomenclature, classification or diagnostic criteria) known to be associated with aortitis, including: Behçet's disease, Cogan's syndrome, Giant cell arteritis (GCA), Granulomatosis with polyangiitis, Inflammatory bowel disease, Kawasaki disease, Polyarteritis nodosum, Polymyalgia Rheumatica, relapsing polychondritis, Rheumatoid arthritis, Sarcoidosis, Sjogren syndrome, spondyloarthropathies, Systemic lupus erythematosus, and Takayasu's arteritis (TAK).

9a. Please select which of the attributes you consider to be part of the definition of isolated aortitis:

Attribute	Required for definition of isolated aortitis	Irrelevant (ie you do not consider this attribute when deciding whether patient has isolated aortitis)
Have to exclude the presence of a defined systemic inflammatory condition (per existing diagnostic or classification criteria)	☐	☐
Have to exclude the presence of any of the clinical features suggesting a defined systemic inflammatory condition (eg – inflammatory arthritis, weight loss, inflammatory bowel disease)	☐	☐
Inflammatory markers should be normal	☐	☐
Have to exclude the presence of radiographic abnormalities in aortic branch vessels (eg – aneurysms, stenosis, thickening of vessel wall)	☐	☐
Other (please specify)	☐	☐
Other (please specify)	☐	☐

For the purpose of consistency for the remainder of the questionnaire, please define **idiopathic aortitis** as aortitis in the absence of a defined systemic inflammatory condition known to be associated with aortitis. We consider **isolated aortitis** a subset of idiopathic aortitis where abnormalities are limited to the aorta (ie appropriate imaging does not reveal evidence of inflammation in aortic branches.)

Please note, for the remainder of the survey participants were able to scroll over the following terms and a definition was immediately provided for reference: ***"defined systemic inflammatory condition", "idiopathic and/or isolated aortitis", "idiopathic aortitis" and "isolated aortitis"***

SECTION 3: REFERRALS

10. Approximately, how many patients with idiopathic and/or isolated aortitis do you see per year (at least once) in your practice?

☐ 0-1
☐ 2-5
☐ 6-10
☐ 11-15
☐ 16-20
☐ >20

11. Approximately, how many NEW patients with idiopathic and/or isolated aortitis are referred to you per year in your practice?

☐ 0-1
☐ 2-5
☐ 6-10
☐ 11-15
☐ 16-20
☐ >20

12. What specialties refer patients with idiopathic and/or isolated aortitis to your practice?

(please select all that apply)

☐ Vascular or cardiac surgery
☐ Cardiology
☐ Internal medicine
☐ Radiology
☐ Pathology
☐ Other (please specify)_____

SECTION 3: REFERRALS (continued)

13. When being referred a patient with suspected idiopathic and/or isolated aortitis, what are some common reasons for the referral?

(please select all that apply)

- ☐ Pathology is positive for aortitis post-aneurysm or aortic valve repair
- ☐ Incidental finding of vascular abnormalities suggestive of aortitis on any imaging modality
- ☐ Discovery of a thoracic aortic aneurysm in a patient with past history of a defined systemic inflammatory condition known to be associated with aortitis
- ☐ Discovery of a thoracic aortic aneurysm in a patient with systemic symptoms or signs (fever, fatigue, weight loss, elevated inflammatory markers) or with other features of a systemic inflammatory condition (inflammatory arthritis, new onset headache +/- visual changes, rashes, etc.)
- ☐ Other (please specify): _____

SECTION 4: INITIAL WORKUP

When assessing a patient for suspected **idiopathic and/or isolated aortitis**, please select what you would screen for in your initial visit.

Recall the definition of a **defined systemic inflammatory condition**

14. HISTORY AND PHYSICAL EXAM:

	Always	Sometimes	Never
Symptoms (eg – limb claudication) or signs (peripheral pulses, blood pressure) that suggest involvement of aortic branch vessels	☐	☐	☐
Symptoms or signs that suggest a defined systemic inflammatory condition (again, make the term clickable)	☐	☐	☐
Infectious symptoms, signs or risk factors (ex: TB, syphilis, HIV, hepatitis)	☐	☐	☐
Constitutional symptoms	☐	☐	☐
History or physical evidence of or risk factors for atherosclerotic vascular disease (diabetes, smoking, hyperlipidemia, hypertension, etc)	☐	☐	☐
Past history of a defined systemic inflammatory condition	☐	☐	☐
Family history of a defined systemic inflammatory condition	☐	☐	☐
Family history of atherosclerotic vascular disease	☐	☐	☐
Other (specify and rank)	☐	☐	☐
Other (specify and rank)	☐	☐	☐

SECTION 4: INITIAL WORKUP (continued)

When assessing a patient for suspected **idiopathic and/or isolated aortitis**, please select what you would screen for in your initial visit.

Recall the definition of a **defined systemic inflammatory condition**

15. LABORATORY INVESTIGATIONS:

	Always	Sometimes	If symptoms or signs present	Never
Complete blood count	☐	☐	☐	☐
Liver function test	☐	☐	☐	☐
Renal function test	☐	☐	☐	☐
C-reactive protein (CRP)	☐	☐	☐	☐
ESR	☐	☐	☐	☐
Infectious workup				
Syphylis screen	☐	☐	☐	☐
TB skin test	☐	☐	☐	☐
HIV serology	☐	☐	☐	☐
Hepatitis serologies	☐	☐	☐	☐
Systemic inflammatory workup				
ANA	☐	☐	☐	☐
ENA panel	☐	☐	☐	☐
dsDNA	☐	☐	☐	☐
Antiphospholipid antibodies	☐	☐	☐	☐
Complements	☐	☐	☐	☐
RF	☐	☐	☐	☐
anti-CCP	☐	☐	☐	☐
ANCA	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐

SECTION 4: INITIAL WORKUP (continued)

When assessing a patient for suspected **idiopathic and/or isolated aortitis**, please select what you would screen for in your initial visit.

Recall the definition of a **defined systemic inflammatory condition**

16. IMAGING:

Imaging modality	Always	Sometimes	Only if suspect abnormalities	Never
Chest X Ray	☐	☐	☐	☐
Chest CT (Angiogram)	☐	☐	☐	☐
Chest MRI (Angiogram)	☐	☐	☐	☐
Echocardiogram	☐	☐	☐	☐
Abdominal ultrasound	☐	☐	☐	☐
Abdominal CT (Angiogram)	☐	☐	☐	☐
Abdominal MR (Angiogram)	☐	☐	☐	☐
Conventional Angiogram	☐	☐	☐	☐
PET	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐

SECTION 4: INITIAL WORKUP (continued)

17. Do you perform temporal artery biopsies in patients with suspected idiopathic and/or isolated aortitis?

☐ Always
☐ Only if symptoms of giant cell arteritis
☐ If symptoms of GCA, systemic symptoms, or elevated inflammatory markers
☐ No

18. Do you usually refer your patients with idiopathic and/or isolated aortitis to a specialized vasculitis treatment centre or expert physician for vasculitis (at least once)?

☐ Not applicable – I am the referral expert physician for a vasculitis centre/clinic
☐ I don't have access to a specialized vasculitis treatment centre
☐ I refer most of my idiopathic and/or isolated aortitis patients for potential transfer of care
☐ I refer most of my idiopathic and/or isolated aortitis patients for second opinion but I continue managing the patients
☐ I refer difficult cases only (for second opinion and/or transfer of care)
☐ I have not referred my patients but have sought advice over the phone
☐ I have never had to seek advice for idiopathic and/or isolated aortitis patients

19. Do you refer patients whom you have diagnosed with idiopathic and/or isolated aortitis to cardiac or vascular surgeons, if a surgeon is not already involved?

☐ Always
☐ If I feel an intervention may be appropriate
☐ No

SECTION 5: TREATMENT

Note: we will ask 4 sets of the same questions for 4 brief clinical scenarios (scenario 1A and B and scenario 2A and B). For each of the scenarios, you may assume that detailed evaluation did not reveal presence of a **defined systemic inflammatory condition**.

Scenario 1: Patient diagnosed with aortitis ON IMAGING (i.e. the involved aorta was not surgically resected)

Scenario 1A: Performed imaging suggested additional involvement of aortic branch vessels (stenoses, aneurysms, or thickening of the wall)

20. On average, how often do you treat such patients with corticosteroids and/or other immunosuppressive agents?

- ☐ Always (or almost always)
- ☐ More often than not
- ☐ About half the time
- ☐ Sometimes
- ☐ Never (or very rarely)

21. For Scenario 1A, will you treat your ASYMPTOMATIC patient with corticosteroids and/or other immunosuppressive agents if they have:

Normal inflammatory markers	□yes	□no
Mildly elevated inflammatory markers	□yes	□no
Markedly elevated inflammatory markers	□yes	□no

Scenario 1B: Performed imaging does not reveal additional involvement of aortic branch vessels (i.e. this is isolated aortitis)

22. On average, how often do you treat such patients with corticosteroids and/or other immunosuppressive agents?

- ☐ Always (or almost always)
- ☐ More often than not
- ☐ About half the time
- ☐ Sometimes
- ☐ Never (or very rarely)

23. For Scenario 1B, will you treat your ASYMPTOMATIC patient with corticosteroids and/or other immunosuppressive agents if they have:

Normal inflammatory markers	□yes	□no
Mildly elevated inflammatory markers	□yes	□no
Markedly elevated inflammatory markers	□yes	□no

SECTION 5: TREATMENT (continued)

24. For Scenario 1 (A or B), if you do place your patient on corticosteroids and/or other immunosuppressive therapy, how long do you aim to continue therapy?

- ☐ ≤ 1 month
- ☐ Between 1 and 3 months
- ☐ Between 3 and 6 months
- ☐ Between 6 and 12 months
- ☐ Between 12 and 24 months
- ☐ Greater than 24 months
- ☐ N/A (you never treat such patients with immunosuppressive therapy)

SECTION 5: TREATMENT (continued)

<u>Scenario 2:</u> patient diagnosed with aortitis ON PATHOLOGY (ie: area of aortitis was surgically removed)

<u>Scenario 2A:</u> Performed imaging suggested additional involvement of aorta or aortic branch vessels (stenoses, aneurysms or wall thickening)

25. On average, how often do you treat such patients with corticosteroids and/or other immunosuppressive therapy?

☐ Always (or almost always)
☐ More often than not
☐ About half the time
☐ Sometimes
☐ Never (or very rarely)

26. For Scenario 2A, will you treat your ASYMPTOMATIC patient with corticosteroids and/or immunosuppressive agents if they have:

Normal inflammatory markers □yes □no

Mildly elevated inflammatory markers □yes □no

Markedly elevated inflammatory markers □yes □no

<u>Scenario 2B:</u> Performed imaging does not reveal additional involvement of aorta or aortic branch vessels (i.e. this is isolated aortitis with all of the involved aorta surgically removed)

27. On average, how often do you treat such patients with corticosteroids and/or other immunosuppressive therapy?

☐ Always (or almost always)
☐ More often than not
☐ About half the time
☐ Sometimes
☐ Never (or very rarely)

28. For Scenario 2B, will you treat your ASYMPTOMATIC patient with corticosteroids and/or other immunosuppressive agents (select all that apply) if they have:

Normal inflammatory markers □yes □no

Mildly elevated inflammatory markers □yes □no

Markedly elevated inflammatory markers □yes □no

SECTION 5: TREATMENT (continued)

29. For Scenario 2 (A or B), if you do place your patient on corticosteroids and/or immunosuppressive therapy, how long do you aim to continue therapy?

☐ ≤ 1 month
☐ Between 1 and 3 months
☐ Between 3 and 6 months
☐ Between 6 and 12 months
☐ Between 12 and 24 months
☐ Greater than 24 months
☐ N/A (you never treat such patients with immunosuppressive therapy)

30. What immunosuppressive agents would you prescribe to patients whom you decide to treat for idiopathic or isolated aortitis?

Immunosuppresive agent	Always	Most often	Less often	Seldom	Never
Corticosteroids	☐	☐	☐	☐	☐
Methotrexate	☐	☐	☐	☐	☐
Azathioprine	☐	☐	☐	☐	☐
Leflunomide	☐	☐	☐	☐	☐
Cyclophosphamide	☐	☐	☐	☐	☐
Mycophenolate mofetil	☐	☐	☐	☐	☐
Tocilizumab	☐	☐	☐	☐	☐
Anti-TNF agent	☐	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐	☐

SECTION 6: FOLLOW UP/MONITORING

Here we will ask you the same set of questions for 2 different scenarios. Again, for both scenarios you may assume the detailed clinical assessment did not reveal presence of a **systemic inflammatory condition**.

Scenario 1: Aortitis diagnosed on pathology (i.e. involved area was resected), absence of additional radiographic involvement of aorta or its main branches and absence of symptoms or signs of systemic inflammation (i.e. this is isolated aortitis)

31. On average, how often are you seeing such patients in follow up in the 1st 2 years after diagnosis?

☐ Every 3 months
☐ Every 6 months
☐ Every 12 months
☐ I do not see them in follow up

32. Which of the following tests do you routinely order to monitor such patients and how often?

	Never	Every 3 months	Every 6 months	Every 12 months	Every 24 months
CBC	☐	☐	☐	☐	☐
Creatinine	☐	☐	☐	☐	☐
ESR	☐	☐	☐	☐	☐
CRP	☐	☐	☐	☐	☐
CXR	☐	☐	☐	☐	☐
Echocardiogram	☐	☐	☐	☐	☐
CT(angiogram) or MR(angiogram) of thorax	☐	☐	☐	☐	☐
CT(angiogram) or MR(angiogram) of abdomen	☐	☐	☐	☐	☐
Abdominal ultrasound	☐	☐	☐	☐	☐
PET	☐	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐	☐

SECTION 6: FOLLOW UP/MONITORING (continued)

33. When (if ever) do you think it's appropriate to stop following such patients provided none of your subsequent investigations reveal any abnormalities?

- ☐ After 1 year
- ☐ After 2 years
- ☐ After 5 years
- ☐ After 10 years
- ☐ These patients should be followed for life

<u>Scenario 2:</u> Aortitis diagnosed on pathology or imaging with remaining areas of radiographic involvement of aorta and/or its main branches

34. On average, how often are you seeing such patients in follow up in the 1st 2 years after diagnosis?

- ☐ Every 3 months
- ☐ Every 6 months
- ☐ Every 12 months
- ☐ I do not see them in follow up

35. Which of the following tests do you routinely order to monitor such patients and how often?

	Never	Every 3 months	Every 6 months	Every 12 months	Every 24 months
CBC	☐	☐	☐	☐	☐
Creatinine	☐	☐	☐	☐	☐
ESR	☐	☐	☐	☐	☐
CRP	☐	☐	☐	☐	☐
CXR	☐	☐	☐	☐	☐
Echocardiogram	☐	☐	☐	☐	☐
CT(angiogram) or MR(angiogram) of thorax	☐	☐	☐	☐	☐
CT(angiogram) or MR(angiogram) of abdomen	☐	☐	☐	☐	☐
Abdominal ultrasound	☐	☐	☐	☐	☐
PET	☐	☐	☐	☐	☐
Other (specify and rank)	☐	☐	☐	☐	☐

SECTION 6: FOLLOW UP/MONITORING (continued)

The following questions pertain to both of the aforementioned clinical scenarios:

36. For patients with idiopathic and /or isolated aortitis whom you decide not to treat with corticosteroids or other immunosuppressive agents at the time of diagnosis, how often do you end up treating them at a later time?

☐ Always or almost always
☐ More often than not
☐ About half the time
☐ Sometimes
☐ Very rarely or never

37. For patients initially diagnosed with idiopathic and /or isolated aortitis, how often do you end up diagnosing them with a defined systemic inflammatory condition at a later time?

☐ Always or almost always
☐ More often than not
☐ About half the time
☐ Sometimes
☐ Very rarely or never

38. In general, how comfortable are you managing patients with idiopathic and/or isolated aortitis?

☐ Perfectly comfortable
☐ Reasonably comfortable
☐ Somewhat uncomfortable
☐ Very uncomfortable

39. Do you feel developing recommendations for management of patients with idiopathic and isolated aortitis would be beneficial?

☐ Yes
☐ No

Comments

Type here

Thank you for participating in our survey. This has assisted us greatly in trying to determine the current practice patterns of Canadian Rheumatologists with respect to patients with idiopathic aortitis. We truly value the information you have provided.