Correction to: Etiology, Diagnosis and Management of Aortitis

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In the original article, the section “Fact Sheet” was not published. This section should give the reader an overview on the most important take-home messages on aortitis. Please see below the missing section.

Fact Sheet

Ten most important points of the inflammatory aortic disease:

1. Aortitis refers to the presence of inflammatory changes in the aortic wall. It may be of infectious origin, but is more frequently of non-infectious etiology, including immunological or connective tissue disorders.
2. The two most frequent causes of aortitis include giant-cell arteritis (GCA) and Takayasu arteritis (TA). These show significant overlap in pathological findings although their epidemiological patterns and clinical presentations are distinct.

3. Temporal artery biopsy, considered the gold standard for diagnosis of GCA, may be false negative in one-third cases as GCA involving the aorta typically does not involve the temporal arteries, resulting in false-negative tissue diagnosis.
4. The clinical course of TA is predominantly divided into two distinct phases: an early acute stage characterized by constitutional symptoms and the late chronic stage characterized by clinical symptoms secondary to stenosis, occlusion or dilatation with less prominent constitutional symptoms.
5. While erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) are usually markedly elevated in patients with GCA, they are unreliable in predicting disease activity in TA patients as factors other than active disease can also cause an elevation of these markers. Elevated ESR values are seen in only 72% of patients during active disease, while ESR values are normal in only 56% of patients with disease remission.
6. Normal aorta is resistant to infection; however, an abnormal aortic wall, such as that associated with atherosclerosis, aneurysm, post-device placement or post-surgery, makes it more susceptible to infection. The infection may be secondary to spread from adjacent sites of infection, by hematogenous route or iatrogenic secondary to an intravascular procedure.
7. Anti-inflammatory therapy with steroids and immunosuppressive drugs is the mainstay of treatment for control of disease activity in non-infectious aortitis. Surveillance for relapse of active disease with periodic comprehensive evaluation of clinical, biochemical and imaging markers of activity is imperative as relapses are common and adversely

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impact the success rates of endovascular and surgical treatment.

8. Aortic steno-occlusive disease has been treated with percutaneous transluminal angioplasty (PTA) successfully, although the results are sparsely reported. Elective stenting is usually avoided as the disease is often long segment requiring stent lengths of 8–10 cm or more; risk of reactivation of underlying disease substantially increasing the in-stent and adjacent segment restenosis; as well as the young age of the patients. Whereas the adjacent segments of aorta grow with time, the stented segment does not, resulting in hemodynamic issues in later life. Also, the loss of pulsatility in long stented segments contributes to adverse outcomes.
9. Role of newer technologies, including drug eluting stents and cutting balloons, is largely unexplored in this clinical setting.
10. Management of infectious aortitis is mainly surgical. Role of endovascular techniques is reserved for bail-out situations in extremely sick patients where surgery is not immediately possible and endovascular techniques are used as a bridge to subsequent surgery after the patient's clinical condition has stabilized.

Five most important numbers of inflammatory aortitis:

1. GCA is the most common form of aortitis in North America, accounting for > 75% of cases.
2. GCA is usually seen in patients over 50 years old with the incidence peaking in the eighth decade of life. It has an incidence of 18.8/100,000 individuals of > 50 years of age per year with a female predilection.
3. TA is the most common cause of aortitis in the tropics, South and Southeast Asia and Japan. It more commonly affects young women (ten times more common than in men) with a peak incidence seen in third decade of life. As many as 31% patients are < 16 years of age in north India.
4. Presence of ≥ 3 American College of Rheumatology criteria (ACR) for GCA has been shown to have a sensitivity of 94% and a specificity of 91% for its diagnosis. Similarly, presence of ≥ 3 ACR criteria for TA has a sensitivity of 91% and a specificity of 98% for its diagnosis.
5. Use of FDG-PET or hybrid imaging with PET-CT or PET-MR has a sensitivity of 60–90% and a specificity

ranging from 88 to 100% for the diagnosis of active inflammation in arteritis.

Three major pivotal studies in last 5 years:

1. Barra L, Kanji T, Malette J, Pagnoux C; CanVasc. Imaging modalities for the diagnosis and disease activity assessment of Takayasu's arteritis: A systematic review and meta-analysis. *Autoimmun Rev.* 2018 Feb;17(2):175–187.
2. Águeda AF, Monti S, Luqmani RA, Buttgerit F, Cid M, Dasgupta B, et al. Management of Takayasu arteritis: a systematic literature review informing the 2018 update of the EULAR recommendation for the management of large-vessel vasculitis. *RMD Open.* 2019 Sep 23;5(2):e001020.
3. de Boysson H, Daumas A, Vautier M, Parienti JJ, Liozon E, Lambert M, et al. Large-vessel involvement and aortic dilation in giant-cell arteritis. A multicenter study of 549 patients. *Autoimmun Rev.* 2018 Apr;17(4):391–398.

Three messages about inflammatory aortitis include:

1. Given the nebulous symptoms and signs and considerable overlap of biochemical and imaging markers, a high clinical index of suspicion and a comprehensive assessment of the clinical picture with laboratory and imaging findings is necessary for an early diagnosis.
2. Assessment of disease activity is the key to planning management, and markers of activity can be clinical, biochemical or imaging. Disease activity in aortitis is poorly understood and often misdiagnosed, an issue that adversely impacts the long-term outcomes of endovascular or surgical treatment.
3. Whereas GCA is characterized by skipped areas of involvement, TA typically has contiguous disease as shown by IVUS studies; some areas are abnormal at both angiography and IVUS; intervening segments may show abnormal IVUS appearances but no visible luminal abnormalities at angiography. This has major therapeutic implications as stents, balloons or surgical grafts placed in angiographically normal, but IVUS abnormal areas have serious issues related to an early or late treatment failure, as evidenced by device induced dissection, restenosis and aneurysm formation at treatment site.

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