# A Review of the Diagnostic Criteria and Management of Takayasu Arteritis

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## Introduction

Takayasu Arteritis (TA) is a large vessel vasculitis mainly affecting the aorta and its branches. Currently, there are no ideal diagnostic studies in detecting TA in a patient with active or inactive disease. Diagnosing TA remains a universal challenge as most diagnoses are made once the disease has progressed or displays an extensive pathological symptom. Various forms of criteria have been made for the diagnosis of TA, each of which is of interest for further review of their efficacies. Namely the Ishikawa criteria and the modified Ishikawa criteria given by Sharma et al. These criteria contain values for laboratory studies, imaging studies, and symptomatology of varying grades. Our study aims to collect a comprehensive review of previous research articles assessing the power of current criteria guidelines for the diagnosis and management guidelines of TA.

## Methods

PubMed was screened for relevant articles using the MeSH search term for Takayasu arteritis diagnosis with pertinent information related to the use of computed tomography (CT), magnetic resonance angiography (MRA), positron emission tomography (PET), ultrasound (US), combined PET/CT, and infrared tomography (IT) from 2000 to 2022. Prevalence of presenting conditions was investigated between these imaging modalities, lab values, and symptoms. Additional specific searches for Ishikawa criteria and the Modified Ishikawa criteria were also investigated amongst the literature review.

Following exclusion and inclusion criteria, 19 of 1282 articles were used in this analysis. Exclusion criteria was placed on papers that did not contain sufficient imaging data such as sensitivities or specificities, additionally if papers did not include symptoms that matched with the Ishikawa criteria, those papers were omitted. Inclusion criteria consisted of papers that had multiple symptoms/major criteria included in its data along with accompanied imaging studies.

## Methods, continued

The Ishikawa criteria consisted of age less than 40 years, two major criteria of left and right subclavian artery lesions and nine minor criteria including high ESR, hypertension, and image proven demonstration of lesions of different arteries. Additionally, the presence of two major criteria or one major plus two or more minor criteria, or four or more minor criteria suggests a high probability of TA. When taking into account the modified Ishikawa criteria, the difference is the removal of the obligatory criteria of age, the signs and symptoms being included in the major criteria, the removal of age from defining hypertension, deletion of the absence of aorto-iliac lesion from the ninth minor criteria and the addition of a tenth minor criteria which consists of coronary artery lesion in patients younger than 30 in the who have no risk factors such as hyperlipidemia, diabetes mellitus or any other known risk factor.

## Results

| Presentation within Literature (n = 15) | Prevalence (n, %) |
|---|---|
| **Vascular findings (n = 6)** | |
| Left subclavian artery involvement | 5 (83%) |
| Right subclavian artery involvement | 3 (50%) |
| Pulmonary artery involvement | 1 (17%) |
| Left common carotid involvement | 4 (67%) |
| Right common carotid involvement | 1 (17%) |
| Brachiocephalic trunk involvement | 2 (33%) |
| Descending thoracic aorta involvement | 2 (33%) |
| Abdominal aorta involvement | 5 (83%) |
| Coronary artery involvement | 1 (17%) |
| Renal artery involvement | 2 (33%) |
| Aortic regurgitation | 0 (0%) |
| **Symptoms (n = 5)** | |
| Fever | 3 (60%) |
| Carotid artery tenderness | 2 (40%) |
| Limb claudication | 2 (40%) |
| Pulselessness | 2 (40%) |
| Pulse differences between limbs | 2 (40%) |
| Hypertension | 2 (40%) |
| Headache | 2 (40%) |
| Dyspnea | 2 (40%) |
| Syncope | 1 (20%) |
| Neck pain | 1 (20%) |
| Changes in vision | 1 (20%) |
| **Labs (n = 7)** | |
| High ESR | 5 (71%) |
| High CRP | 4 (57%) |
| Elevated PTX-3 | 1 (14%) |

Table 1. Most common presentations of Takayasu arteritis in the literature

## Results, continued

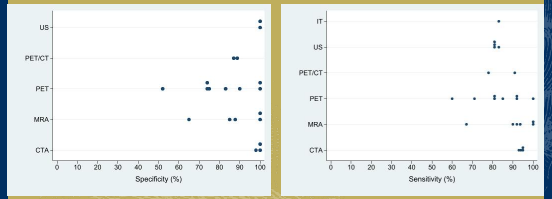


Figure 1. Reported specificities (n = 17) and sensitivities (n = 25) of diagnostic imaging methods for Takayasu arteritis in the literature

The combined data shows strong sensitivity and specificity in the diagnosis of TA with the use of CTA both in inactive and active disease, however, MRA shows increased sensitivity and specificity. In terms of laboratory studies, various biomarkers such as C-reactive protein and erythrocyte sedimentation rate showed variable significance in its sensitivity and specificity, albeit useful for favoring large vessel vasculitis. Pentraxin-3 is an additional biomarker that has shown increase in the disease process of TA. Overall, the diagnosis results in similar treatment plans including the use of corticosteroids, TNF-α inhibitors such as Infliximab or the IL-6 receptor inhibitor, Tocilizumab.

## Conclusions

After reviewing the most recent literature, it is evident that the modified Ishikawa criteria paired with CTA or MRA can provide diagnostic yields with higher sensitivities and specificities. Despite the higher diagnostic yield, there is still a need for further investigation on more sensitive and specific biomarkers and imaging studies in identifying TA in earlier stages.

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