Takayasu Arteritis: Diagnosis, Treatment and Prognosis

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Takayasu Arteritis: Diagnosis, Treatment and Prognosis

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Takayasu arteritis (TA) is a chronic nonspecific granulomatous vasculitis affecting aorta and its main branches, coronary and pulmonary arteries. TA often occurs in young women and has a characteristic heterogeneity depending on ethnicity and geographical location. Although the pathogenesis of TA remains unclear, the interaction of many factors, such as autoimmunity, inflammation, genetic and environmental factors and so on, is involved in the occurrence and development of TA. Angiography, which is recognized as the gold standard in evaluating vascular lesions in TA, combined with computer tomography angiography (CTA), magnetic resonance angiography (MRA), ultrasonography, $^{18}$Fluorodeoxyglucose positron emission tomography ($^{18}$F-FDG-PET) could not only provide important information for early diagnosis but also detect disease activity, and thus further guide the treatment in TA. In addition, beside the commonly used corticosteroids, immunosuppressive agents, percutaneous transluminal angioplasty (PTA) and surgical revascularization, anti-tumor necrosis factor (TNF) agent has been more widely used in refractory cases of TA. The objective of this review is to systemically describe the pathogenesis, clinical characteristics, diagnosis, treatment and prognosis of TA.

Keywords diagnosis, disease activity, prognosis, Takayasu arteritis, treatment

INTRODUCTION

Takayasu arteritis (TA), which is also known as pulseless disease, aortitis syndrome, aortic arch syndrome, and atypical coarctation of aorta, is a chronic nonspecific granulomatous vasculitis of unknown etiology affecting aorta and its main branches, coronary and pulmonary arteries [1–4]. TA usually occurs in young women with atypical early symptoms and has a growing trend in recent years. The pathogenesis of TA is complex, involving genetic factors, cellular and humoral immunity, infection, sex hormone, and environmental factors and so on. Great progress has been made in the diagnosis and treatment of TA currently. At present, the prognosis of TA has been improved and the long-term survival rate has been increasing. In this review, we systemically describe the pathogenesis, clinical characteristics, diagnosis, treatment and prognosis of TA.

PATHOGENESIS

The pathogenesis of TA is multifactorial and remains unclear. Autoimmunity, endocrine abnormalities, genetic and environmental factors, and inflammation are...
Recent progress in TA

considered to play important roles in the pathogenesis of this disease [5, 6–10]. Cell-mediated autoimmunity mechanisms in vessel lesions associated with TA have been recently elucidated. T-cell-dependent immunity, chemokine- and cytokine-dependent immunity, and B-cell-dependent vascular inflammation were thought to be the main pathological mechanisms resulting in the arterial wall injury involved in TA [2].

Elevated levels of HLA-DR+, CD28+, CD45RA+, and CD45RO+ were reported in TA. Moreover, CD4+ expansions and increased ratio of CD4+/CD8+ had some correlations with disease activity. Further, the interaction between dendritic cells and lymphocytes may also contribute to the immune and inflammatory response involved in TA [11, 12]. Higher basal activity of protein kinase C (PKC) and concentrations of intracellular calcium, and lower levels of cyclic adenosine monophosphate (cAMP) were observed indicating the activation of PKC-calcium pathway in patients with TA [13]. In addition, restricted usage of T-cell receptor Vα as well as Vβ genes in infiltrating cells was observed in TA showing that a specific antigen in the aortic tissue was targeted [14]. However, previous studies on antibodies in TA showed conflicting findings regarding the involvement of B cell immunity in the pathogenesis of the disease, an aspect that remains controversial [6].

Generally, the pathologic process is characterized as follows: inflammatory cell infiltration (mainly including γδ+ T cells, cytotoxic T lymphocytes, T helper cells, natural killer cells and macrophages) and cell-mediated immune response affect intima, media and adventitia (media is the mainly involved), which eventually lead to the affected aorta and its main branches thickening, stenosis, occlusion or occasionally result in aortic dissection/aneurysm formation [1, 2, 15, 16].

TA is a kind of chronic inflammatory disease in itself. The course of TA including active and quiescent phases demonstrates different inflammatory states of arterial lesions. Oxidative stress, which is interrelated with inflammation and could induce the expression and production of heat shock proteins (HSP), proinflammatory cytokines and adhesion molecules, has been shown to be correlated with disease activity of TA, and thus involved into the pathogenesis of TA [17]. Moreover, TA had a high rate of atherosclerotic plaques and, thus, promoted the formation and occurrence of atherosclerosis [18, 19]. Many TA patients showed increased prevalence of hypertension, triglycerids levels and other traditional risk factors for cardiovascular disease comparing to healthy controls [20]. All these findings further illustrate the inflammatory mechanisms in the pathogenesis of TA.

Several studies have reported that human leukocyte antigen (HLA)-link genes is associated with the development of TA. Compared with healthy controls, significantly higher increased frequencies of HLA-A10, B39, B52, Bw52, B5, DR2 were confirmed in TA in Japanese population, and increased frequency of HLA-Bw52, Cw6, DR7, DQw2 were observed in Korean patients with TA [21–23]. Moreover, in Mexican population, HLA-B39, B44, B52 alleles had the susceptibility to TA [24]. In Caucasian North American patients with TA, HLA-DR2, DR4 and DQw3 were reported to have no significant positive correlations with TA susceptibility, severity and complications. Furthermore, DR1 antigen was considered to be a protective agent against TA development [25].

Furthermore, HLA is also related to the clinical manifestations in TA. Patients with the haplotype of Bw52-Dw12 showed increased risk of showing active inflammatory state and rapid progression of morbid conditions associated with TA in Japanese [26]. Kasuya et al. also found that TA patients with Bw52 antigen were more prone to have left ventricular dysfunction than those without, and thus demonstrated poor prognosis [27]. In addition, genetic polymorphisms of interleukin (IL)-2, 6, 12, which could be
linked to cytokine expression and production, were considered as susceptibility factors for TA in Turkey [28].

Moreover, endocrine abnormalities have also been considered to be involved into the pathogenesis of TA. TA is more common in young women. Serum estradiol and progesterone levels in patients with TA were significantly higher than those in healthy women. At the same time, the estrogen levels in the urine of women patients with TA during the follicular phase and luteal phase were also significantly increased comparing with normal healthy controls. Moreover, in animal experiments, long-term treatment with estrogen could induce pathological changes in aorta and its branches, similar to TA lesions [29]. These data suggest that increased levels of estrogen is also an important factor in the pathogenesis of TA.

**CLINICAL CHARACTERISTICS**

**Clinical Classification**

There exist three reported criteria for the clinical classification of TA according to the distribution of the lesions. Initially, in 1967, Ueno et al. [30] classified TA into three types: type I, involvement of the aortic arch and its branches; type II, involvement of the descending aorta and abdominal aorta; type III, the combined features of type I and type II. This classification method has some limitations because it does not consider the pulmonary artery involvement. Accordingly, in 1977, Lupi-Herrera et al. [31] proposed type IV based on Ueno classification showing that type IV was considered when pulmonary artery was involved regardless of the aortic involvement. Next, Hata et al. [32] provided another classification that is listed as follows: type I, involvement of the main branches from the aortic arch; type IIa, involvement of the ascending aorta, aortic arch and its branches; type IIb, involvement of the ascending aorta, aortic arch and its branches, and thoracic descending aorta; type III, involvement of the thoracic descending aorta, abdominal aorta and/or renal arteries; type IV, involvement of the abdominal aorta and/or renal arteries; and type V, the combined features of type IIb and IV.

Aorta involvement also shows regional difference indicating that ethnic factors may play an important role in the pathogenesis of TA. For example, ascending aorta is often affected in Japan, while in other Asian countries, thoracic and abdominal aortas are usually involved [1]. In China, type I was the most common, followed by type V and IV. Type I is more often in adult patients than in pediatric patients. The mechanisms of this age-related difference remain unclear and need further studies to elaborate [33].

Coronary artery involvement, which is often affected the ostial of left main coronary artery, is observed in about 10–30% of the patients with TA [3], whereas renal artery was reported in approximately 50% of TA cases [4]. Pulmonary artery involvement, which is a TA-specific pathology, is often underestimated because pulmonary angiography is not a clinical routine examination. It was reported that abnormal pulmonary angiography in patients with TA is about 30–74% [33].

**Activity of TA**

**Criteria for disease activity**

The natural course of TA consists of active and quiescent phases. The commonly used criteria to evaluate disease activity are the National Institute Health (NIH) criteria including: (1) systemic features such as fever or musculoskeletal problems (no other cause identified); (2) elevated erythrocyte sedimentation rate (ESR); (3) features of vascular ischemia or inflammation, such as claudication, diminished or absent pulse, bruits, vascular pain (carotodynia), asymmetric blood pressure in either upper or
Recent progress in TA lower limbs (or both); and (4) typical angiographic features. Patients who demonstrate new onset or worsening of at least two of the five features listed above are considered as active disease. Accordingly, disease activity in TA could also be described by disease activity score: each of the above listed features is given a score of 1, and a total score of at least 2 defines active disease [16, 36].

Criteria for remission of TA
Remission was defined as the resolution of clinical and laboratory features of active disease and the absence of new vascular lesions, as determined by sequential imaging studies. Improvement or partial remission was considered if the dosage of glucocorticoid or immunosuppressive agents could be reduced by at least 50%, compared with the initial dosage, before the disease recurred. Complete and sustained remission was defined as the absence of features of active disease, the absence of new lesions on sequential imaging studies, and no glucocorticoid or immunosuppressive agent treatment or less than 10 mg/day of glucocorticoid dosage for more than 6 months [38, 39].

Even though the above criteria of TA activity and remission have been conventionally used as reliable measures in clinical setting, histopathological assessment is still the gold standard in evaluating disease activity [40]. There exist some patients who despite their clinical remission status, were diagnosed based on histopathological evidence of inflammation. It was reported that 44% of clinically inactive TA patients showed histologically active disease by surgical bypass biopsy. Moreover, over 60% of the patients considered in prolonged remission by clinical criteria demonstrated new lesions by sequential angiographic evaluation [16, 41, 42].

When the diagnosis of TA is confirmed, evaluating the disease activity is necessary for the treatment. However, the presently used imaging modalities and serological biomarkers in TA lack either sensitivity or specificity. Thus, potential noninvasive, rapid, easy to perform, sensitive and specific tools are necessary in confirming TA activity.

DIAGNOSIS
The diagnosis of TA is based on atypical clinical symptoms and signs, increased concentrations of biomarkers, and confirmed by angiographic morphology findings. In the meanwhile, the establishment of TA should also exclude other similar conditions such as Ehlers–Danlos and Marfan syndrome, tuberculosis, giant cell arteritis, syphilis, spondyloarthopathies, Cogan’s syndrome, Buerger’s, Behcet’s, and Kawasaki disease [4, 43].

Diagnose Approach
At present, imaging techniques, including angiography, computer tomography angiography (CTA), magnetic resonance angiography (MRA), ultrasonography, 18Fluorodeoxyglucose positron emission tomography (18FDG-PET), are increasingly being used in not only diagnosing but also monitoring disease development in TA.

Angiography
Angiography is the gold standard in diagnosing TA, especially for advanced stage of vascular lesions, such as artery stenosis, occlusion, restenosis after dilatation, or aneurysm formation and so on. Angiography could provide reliable evidence for lesion location and extent, and thus guide treatment method for TA. However, angiography shows low diagnostic value in timely detecting the early stage of lesion development. At the same time, angiography in itself is an invasive examination, and
patients need to receive a large dose of radiation. Therefore, angiography is not suitable for long-term follow-up of patients with TA [34, 45].

**CTA**

CTA is reported to be convenient, noninvasive and useful in evaluating vascular lesions. CTA can not only demonstrate the lumen of aorta and its branches but also detect vessel wall lesions. In addition, intravascular blood flow can be fully displayed by CTA. CTA could indicate the length and degree of vascular lesion, and collateral vessel formation in TA as well. Although the disadvantage of CTA is that patients need to receive a large dose of radiation, and it provides little information for early stage of vascular lesion, CTA still shows important clinical value in diagnosing vascular lesion in TA at middle and advanced stages [46, 47].

**MRA**

MRA, which is similar to CRA, has been observed to facilitate diagnosis at the early stage of TA, when there only exists vessel wall thickening or edema. Furthermore, MRA findings are usually positively correlated with ESR and CRP levels [48]. MRA could provide clear images of the vessel wall thickness and lumen structure, especially for lesions in the descending aorta. It has been reported that the diagnostic accuracy of MRA could achieve the same as artery angiography. However, MRA is expensive, and may overestimate the degree of stenosis of branch arteries, and have poor imaging performance in distal branches, or aortic calcification [35, 49].

**Ultrasonography**

Color Doppler ultrasound demonstrates important diagnostic value for patients with TA at early stage of vessel wall inflammation. Moreover, ultrasonography could also show blood flow within the vessel, which can better evaluate the degree of stenosis. At the same time, it could differentiate vasculitis and atherosclerosis. Due to its noninvasive, ultrasonography has been widely used in the early diagnosis, follow-up and long-term monitoring of disease activity in TA. However, the inadequacies in ultrasonography indicate that it cannot provide intuitive and comprehensive morphology change information in vessels, and it is difficult to detect some vascular lesions, such as the subclavian artery, right pulmonary artery, thoracic and abdominal aorta, renal artery and superior mesenteric artery and so on. In addition, the operator’s proficiency may also affect the sensitivity and reliability of ultrasonography results [35, 44, 50].

**18F-FDG-PET**

18F-FDG-PET has been considered as an important examination method for the early diagnosis, monitoring the disease activity, and response to treatment of TA. Several studies reported that 18F-FDG-PET may be a screening method in early atypical TA [43, 51, 52]. 18F-FDG-PET is superior to MRA in monitoring blood vessel wall inflammation in the early stage of TA. This might be due to the former being sensitive to inflammation of the vascular wall, while the latter is prone to identify the blood vessel wall thickening or edema. Moreover, inflammation of the vessel wall is usually occurs earlier than the morphological changes such as thickening or edema [37, 46, 53].

The disadvantages of 18F-FDG-PET are the high costs, incapability in displaying the changes in vessel wall structure and blood flow, and low specificity. For example, in elderly patients, 18F-FDG-PET cannot distinguish between TA and atherosclerosis [54, 55, 60]. Therefore, its clinical application value still needs further studies to confirm.

In summary, the above-mentioned several imaging methods show that angiography is still the gold standard in diagnosing TA. CTA, MRA, ultrasonography and 18F-FDG-PET, which are complementary with angiography, are important auxiliary.
examination methods. MRA, ultrasonography and $^{18}$F-FDG-PET could found early vessel wall inflammation and structural changes in TA, while CTA is suitable for diagnosing TA at middle and advanced stages. The combination of the above-listed methods could not only help early diagnosis but also be able to determine the disease activity, and then guide treatment in TA.

**Diagnostic Criteria**

The commonly used diagnostic criteria of TA in adults, which were proposed by the American College of Rheumatology in 1990, were listed as follows: age of onset $\leq$ 40 years, limb claudication, decreased branchial artery pulse, the difference in bilateral limb systolic blood pressure $> 10$ mmHg, a bruit over the subclavian arteries or the aorta, abdominal aortic murmur, abnormal arteriography indicating narrowing or occlusion of the entire aorta and its main branches [56]. The presence of three or more of the six criteria showed a sensitivity of 90.5% and a specificity of 97.8% in diagnosing TA.

In addition, Ishikawa et al. proposed Ishikawa diagnostic criteria in 1988, and then, Sharma et al. [57] revised these criteria as clarified in Table 1, with the sensitivity 96% and specificity 96%. The presence of two major or one major and two minor criteria or four minor criteria may demonstrate a high probability of TA.

**TREATMENT**

Medical treatment (including corticosteroids, immunosuppressive agents and anti-TNF agents), percutaneous transluminal angioplasty (PTA) and surgical vascular reconstruction are currently used in the treatment of TA.

**Medical Treatment**

Immune mechanisms have been confirmed to be involved in the destruction of the vessel wall in TA. At the same time, administration of corticosteroids and other immunosuppressive agents has demonstrated positive anti-inflammatory effects in a majority of patients with TA. Glucocorticoid is the first-line medical treatment in order to achieve remission, especially in active stage of TA. The medication starts with larger doses, continuing with maintenance doses until remission. It was reported that glucocorticoid treatment success rate ranged from 20% to 100% [33, 58]. Only approximately 60–80% of patients who used corticosteroids only were cured. However, over 50% of patients who achieved remission relapsed during dose tapering [59]. Moreover, HLA haplotype is associated with not only the pathogenesis but also the response to steroid treatment in TA. Patients with HLA A24-B52-DR2 haplotype are prone to accelerated inflammatory progression and tend to resist steroid therapy comparing to those without [1].

Immunosuppressive drugs, such as mycophenolate mofetil, methotrexate, azathioprine, cyclophosphamide, are usually added due to glucocorticoid resistance, or relapse during glucocorticoid dose reduction, or serious side effects associated with steroids treatment. Repeated and prolonged courses of immunosuppressive therapy are often required due to the relapsing nature of TA.

An increasing number of observations indicated that TNF-alpha blockade therapy, infliximab, was effective in patients with TA who were resistant to the common medical therapy including glucocorticoids and immunosuppressants, and may become a possible treatment method for refractory TA cases [39, 61, 62].

In all, the general treatment strategy of TA patients could be summarized as follows: initially, glucocorticoid is used with the usual dose of 1 mg/kg/day, and the dosage is
TABLE 1 Revised Ishikawa diagnostic criteria for TA

<table>
<thead>
<tr>
<th>Major diagnostic criteria</th>
<th>Clinical definition</th>
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<tbody>
<tr>
<td>(1) Left mid subclavian artery lesion</td>
<td>The most severe stenosis or occlusion present in the mid portion from the point 1 cm proximal to the vertebral artery orifice up to that 3 cm distal to the orifice determined by angiography</td>
</tr>
<tr>
<td>(2) Right mid subclavian artery lesion</td>
<td>The most severe stenosis or occlusion present in the mid portion from the right vertebral artery orifice to the point 3 cm distal to orifice determined by angiography</td>
</tr>
<tr>
<td>(3) Typical signs and symptoms (at least 1 month duration)</td>
<td>Limb claudication, pulselessness or uneven pulse in limbs, undetectable blood pressure or the difference in bilateral limb systolic blood pressure &gt;10 mmHg, fever, neck pain, transient amaurosis, blurred vision, syncope, palpitations, dyspnea</td>
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</table>

<table>
<thead>
<tr>
<th>Minor diagnostic criteria</th>
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<tbody>
<tr>
<td>(1) Elevated ESR</td>
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<tr>
<td>(2) Carotid artery tenderness</td>
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<tr>
<td>(3) Hypertension</td>
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<tr>
<td>(4) Aortic regurgitation or dilatation</td>
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<td>(5) Pulmonary artery lesion</td>
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<td>(6) Left mid common carotid lesion</td>
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<tr>
<td>(7) Distal brachiocephalic trunk lesion</td>
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<tr>
<td>(8) Descending thoracic aorta lesion</td>
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<tr>
<td>(9) Abdominal aorta lesion</td>
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<tr>
<td>(10) Coronary artery lesion</td>
</tr>
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</table>

slowly reduced according to the disease activity; then, immunosuppressive agents are used when glucocorticoid-resistance, or relapse during glucocorticoid dose reduction occurred; finally, anti-TNF therapy may be performed for those refractory patients.

PTA
When significant hemodynamic disorders occur because of vascular stenosis and occlusion, PTA or vascular reconstruction is necessary. Research data showed that the indications for PTA were more than 70% of the normal diameter or a hemodynamically significant aortic narrowing (peak systolic gradient of more than 50 mm across the stenotic aortic lesion), and more than 20 mmHg of pressure gradient across the stenosis, and clinically inactive stage of TA [42, 63].

However, restenosis after PTA with or without drug-eluting stent, which is unavoidable, is still a concern. Previous studies have reported that PTA showed high rate of initial success (about 89%) and patency (about 79%) during follow-up (43 months) [33, 58]. Restenosis occurred in 77.3% of the procedures that were initially successful [33, 36, 38]. The possible explanation for restenosis might be that the vessel lesions
in TA are usually long, fibrotic, and almost completely occluded. In the meanwhile, PTA in itself, is a kind of vascular injury, which could promotes vascular endothelial cell proliferation and stenosis. PTA with or without stenting could just only treat the superficial results while not deal with the disease itself.

**Surgical Vascular Reconstruction**

It has been reported that performing surgery during an active phase of TA led to increased likelihood of graft rejection and progression of symptomatic disease at other sites [64]. The eligibility criteria for surgery are as follows: hypertension caused by severe renal artery stenosis, limb claudication, progressive aneurysm enlargement, which prone to dissection or rupture, cerebrovascular ischemia or critical stenosis of three or more cerebral vessels, coronary artery ischemia, moderate to severe aortic regurgitation, and severe aortic coarctation [62, 65]. That is to say, patients who had the state of clinical progression and major complications, and thus with a poor prognosis should be considered for surgical therapy [66].

Furthermore, surgical intervention could not only reduce the complications caused by TA but also increase the long-term survival of patients [66, 67]. Surgical therapy of coronary artery involvement in TA showed positive outcome with survival rates of 86.5 ± 7.3% at 5 years and 81.4 ± 8.4% at 10 years [58].

Surgical revascularization showed higher long-term artery patency rate than PTA procedures (the restenosis rate occurred in about 36% of bypass procedures and 78% of angioplasty). The long-term effects of revascularization were better than angioplasty [3, 62]. However, it was also accompanied by more complications [69, 70]. Anastomotic aneurysm was observed to be the usual complication after surgery in TA with a cumulative incidence of 13.8% at 20 years making regular imaging modalities follow-up necessary in monitoring this complication [68].

**SURVIVAL RATE AND PROGNOSIS**

The prognosis of TA is mainly dependent on two aspects: the prolonged state of inflammation progression, and major complications resulted from the vascular lesions [3]. At the same time, consistently elevated ESR also had an important reference value in predicting TA prognosis [66]. The criteria for major complications, proposed by Ishikawa et al. [66], were the presence of at least one of the following conditions due to TA: (1) microaneurysm formation (stage 2 retinopathy), (2) severe hypertension (the brachial pressure of ≥200 mmHg systolic or ≥110 mmHg diastolic; alternatively, the popliteal pressure of ≥230 mmHg systolic or ≥110 mmHg diastolic may be used), (3) grade 3+ or 4+ aortic regurgitation, and (4) angiographic demonstration of an aortic or arterial aneurysm with a diameter more than twice the normal. Patients with two or more of complications, including Takayasu's retinopathy, secondary hypertension, aortic regurgitation, and aortic or arterial aneurysm, were also considered to have major complications even if each of the complication did not meet the above-listed criteria.

The more complications the higher mortality rate, and the presence of serious complications was closely related to poor prognosis. The common causes of death may be acute myocardial infarction, congestive heart failure, cerebrovascular accident, renal failure, postoperative complications and so on [71–73]. The reported survival rate in patients with TA was inconsistent in several studies, ranging from 67 to 100% (at both 5 and 10 years follow up). Ishikawa et al. proposed that the 15-year survival was 66.3% versus 96.4% for patients with and without a major complication, 67.9% versus 92.9% for patients with and without a progressive course, 58.3% versus 92.7% for age >35 years and ≤35 years [66]. Later, Miyata et al. also reported that the survival rate after surgical treatment in TA was 73.5% at 20 years and it was closely related to the age.
at surgery. Patients over 35 years presented a 2.74-fold higher risk of death compared to those less than 35 years [67].

CONCLUSIONS

TA is a chronic nonspecific inflammatory disease involving the aorta and its major branches, coronary and pulmonary arteries. It often occurs mainly in young women, and has geographical and ethnic distribution difference. Its clinical manifestations demonstrate diversity in different gender and ethnic populations. Autoimmune, inflammatory, genetic, environmental, endocrine factors have some relationships with the occurrence and development of TA. Angiography has been considered to be the gold standard in diagnosing TA. The combination of angiography and other diagnostic techniques, including CTA, MRA, ultrasonography, $^{18}$F-FDG-PET, could not only be useful for early diagnosis but also provide key information regarding the state of disease activity, and then guide treatment.

At present, anti-TNF agents have been widely used in refractory TA, in addition to glucocorticoids and immunosuppressive agents. In order to improve long-term outcome, PTA with or without stent placement and revascularization procedures should not be performed until patients are in remission stage when inflammation is controlled effectively. Timely and regular treatment and long-term follow-up are necessary in order to relieve stenosis and restenosis, reduce the incidence of complications and improve long-term survival.

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Declaration of Interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

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