Preethi G N, 2026, 14:3
ISSN (Online): 2348-4098
ISSN (Print): 2395-4752

International Journal of Science, Engineering and Technology
An Open Access Journal

Takayasu Arteritis in Children and Young Adults: Current Perspectives on Pathogenesis, Clinical Features, Diagnosis, Monitoring and Management

Preethi G N[1], Ashwini Angadi[2], Janaki R Torvi[3], Shaista Omer[4], Adarsh GS[5]

[1,4]TMAE Society's MMJG College of Pharmacy, Haveri - 581110, Karnataka, India.
[3]Departments of Pharmacology, Karnataka Medical College and Research Institute, Hubballi-580029, Karnataka, India.
[2,5]Sonia Education Trust's College of Pharmacy, Dharwad-580002, Karnataka, India.

Abstract- Takayasu arteritis (TA) is a rare chronic granulomatous vasculitis involving the aorta and its major branches. The disease predominantly affects young females and may lead to progressive vascular stenosis, occlusion, aneurysm formation, and ischemic complications. Early diagnosis remains challenging because initial symptoms are often nonspecific and laboratory markers may not accurately reflect disease activity. Advances in imaging techniques, immunosuppressive therapy, biologic agents, and vascular interventions have significantly improved patient outcomes. This review summarizes the epidemiology, pathogenesis, clinical manifestations, diagnosis, disease monitoring, treatment strategies, and prognosis of Takayasu arteritis, with special emphasis on pediatric disease.

Keywords: Takayasu arteritis, large-vessel vasculitis, children, diagnosis, biologic therapy, vascular inflammation.

I. INTRODUCTION

Takayasu arteritis (TA) is a chronic inflammatory disease characterized by granulomatous inflammation of the aorta and its major branches, leading to vascular wall thickening, stenosis, occlusion, thrombosis, and aneurysm formation. The disease was first described in 1908 by Japanese ophthalmologist Mikito Takayasu after observing characteristic retinal vascular changes in a young female patient.[1]

The disease has also been referred to as "pulseless disease," "aortic arch syndrome," and "occlusive thromboaortopathy" because of its characteristic involvement of major arteries resulting in absent peripheral pulses.[2,3] Although uncommon, TA remains one of the most important causes of large-vessel vasculitis in children and young adults and can result in significant morbidity and mortality if not diagnosed and treated early.[4]

The disease primarily affects women during the second and third decades of life. Clinical manifestations range from constitutional symptoms to severe vascular complications such as stroke, myocardial ischemia, renovascular hypertension, and heart failure.[2,3]

Epidemiology

Takayasu arteritis occurs worldwide but is more prevalent in Asian countries, India, Latin America, and parts of Africa. The annual incidence is estimated at approximately 1–3 cases per million population.[2,3] Women account for nearly 80–90% of cases. The highest incidence occurs during the second and third decades of life, although childhood-onset disease is increasingly recognized. In pediatric populations, TA is considered the most common large-vessel vasculitis and a significant cause of renovascular hypertension.[4]

Studies have demonstrated geographic variation in disease frequency and clinical presentation. Asian patients frequently present with involvement of the aortic arch and its branches, whereas abdominal aortic and renal artery involvement are more commonly reported in Indian populations.[3]

II. HISTORICAL BACKGROUND

The first description of the disease was made by Mikito Takayasu in 1908 during a meeting of the Japanese Ophthalmology Society. He reported a young woman with characteristic retinal vascular anastomoses. Subsequently, additional patients were identified with absent radial pulses and similar

ocular findings, leading to recognition of the disease as a systemic vasculitis.[1]

Over the years, several classification systems have been proposed based on angiographic findings and clinical manifestations. Ishikawa later introduced a prognostic classification system that categorized patients according to disease complications and severity.[7]

Pathogenesis

The exact etiology of Takayasu arteritis remains unknown. Current evidence suggests that the disease develops through complex interactions among genetic susceptibility, immune dysregulation, and environmental factors.[2]

Genetic Factors

Several genetic associations have been identified, particularly with HLA-B52 and HLA-B5 alleles. Additional susceptibility loci involving IL12B, FCGR2A, FCGR3A, and HLA class II genes have also been reported.[2]

Immunological Mechanisms

Histopathological examination reveals granulomatous inflammation affecting all layers of the arterial wall. Activated T lymphocytes, macrophages, giant cells, and pro-inflammatory cytokines contribute to vascular injury. Chronic inflammation results in fibrosis, intimal proliferation, and luminal narrowing.[1,2]

Infectious Triggers

Although no infectious agent has been conclusively proven to cause TA, Mycobacterium tuberculosis has been extensively studied as a possible trigger. Molecular mimicry between mycobacterial heat-shock proteins and human proteins may induce autoimmune vascular injury in genetically susceptible individuals.[2]

Pathology

The hallmark pathological feature of TA is panarteritis involving the intima, media, and adventitia.

Histological findings include:

- Mononuclear inflammatory infiltrates
- Giant cell formation
- Intimal hyperplasia
- Medial destruction
- Elastic fiber fragmentation
- Fibrosis of the vessel wall

These changes eventually lead to stenosis, occlusion, aneurysm formation, and thrombosis.[1,4]

III. CLINICAL MANIFESTATIONS

The clinical presentation varies depending on disease stage and vascular territories involved.

Early Systemic Phase

The initial inflammatory phase is characterized by nonspecific constitutional symptoms such as:

- Fever
- Malaise
- Fatigue
- Weight loss
- Arthralgia
- Myalgia
- Night sweats

Because these manifestations are nonspecific, diagnosis is often delayed.[3]

Vascular Occlusive Phase

As vascular damage progresses, symptoms related to ischemia develop, including:

- Limb claudication
- Absent or diminished pulses
- Blood pressure discrepancies between limbs
- Carotidynia
- Vascular bruits
- Headache
- Dizziness
- Syncope
- Visual disturbances

Neurological complications result from involvement of carotid and vertebral arteries and may include transient ischemic attacks and stroke.[3,5]

Pediatric Takayasu Arteritis

Childhood-onset TA differs somewhat from adult disease and is associated with substantial disease burden.

Common presenting manifestations include:

- Hypertension
- Constitutional symptoms
- Headache
- Heart failure
- Neurological deficits
- Renal artery stenosis

In a pediatric cohort study, approximately half of affected children experienced disease flares despite treatment, highlighting the aggressive nature of childhood TA.[6]

Vascular Involvement

TA can affect virtually any large artery.

Commonly involved vessels include:

- Aortic arch
- Subclavian arteries
- Common carotid arteries
- Vertebral arteries
- Renal arteries
- Mesenteric arteries
- Coronary arteries
- Pulmonary arteries

The resulting vascular lesions include:

- Stenosis
- Complete occlusion
- Thrombosis
- Aneurysm formation
- Arterial dissection

These lesions may produce ischemic complications involving the brain, kidneys, heart, and extremities.[2,4]

Renal Involvement

Renal artery stenosis is one of the most clinically significant manifestations of TA. Renovascular hypertension is often the first indication of renal involvement and may lead to:

- Resistant hypertension
- Progressive renal dysfunction
- Chronic kidney disease
- End-stage renal disease

The severity of renal damage depends on the extent of vascular involvement and duration of ischemia. Early diagnosis and intervention are essential to preserve renal function.[8]

IV. DIAGNOSTIC EVALUATION

Diagnosis is based on a combination of clinical findings, laboratory investigations, and vascular imaging.

Laboratory Investigations

Common laboratory abnormalities include:

- Elevated ESR
- Elevated CRP
- Normocytic anemia
- Leukocytosis
- Thrombocytosis

However, laboratory markers often fail to accurately reflect disease activity.[5]

Imaging Modalities

Imaging plays a central role in diagnosis and follow-up.

Computed Tomography Angiography (CTA)

CTA provides detailed visualization of vessel wall thickening, stenosis, occlusion, and aneurysms.

Magnetic Resonance Angiography (MRA)

MRA offers excellent assessment of vascular anatomy without radiation exposure.

Doppler Ultrasonography

Useful for evaluating carotid and subclavian artery involvement.

Conventional Angiography

Traditionally considered the gold standard for diagnosis.

Positron Emission Tomography (PET)

Can identify active vascular inflammation before structural damage becomes apparent.[2,3]

Disease Monitoring

Monitoring disease activity remains one of the greatest challenges in TA management.

ESR and CRP are commonly used but have limited sensitivity and specificity. Studies have

demonstrated progression of vascular disease despite normal inflammatory markers.[5]

Therefore, disease assessment should combine:

- Clinical evaluation
- Laboratory parameters
- Serial vascular imaging

Regular imaging is particularly important because vascular progression may occur silently.[3],[5]

V. MEDICAL MANAGEMENT

Corticosteroids

Glucocorticoids remain the cornerstone of treatment and effectively suppress active vascular inflammation in most patients.[3]

Conventional Immunosuppressive Agents

To reduce steroid dependence and maintain remission, additional immunosuppressive medications are frequently used:

- Methotrexate
- Azathioprine
- Cyclophosphamide
- Mycophenolate mofetil
- Leflunomide

These agents are especially useful in relapsing disease.[2],[3]

Biologic Therapy

Recent advances have introduced biologic agents targeting specific inflammatory pathways.

Commonly used biologics include:

- Tumor necrosis factor inhibitors
- Tocilizumab

In pediatric studies, biologic therapies have demonstrated improved flare-free survival and superior disease control compared with conventional therapies.[6]

Surgical and Endovascular Management

Surgical intervention becomes necessary when severe vascular lesions cause critical ischemia or uncontrolled hypertension.

Indications include:

- Severe renal artery stenosis
- Cerebral ischemia
- Critical limb ischemia
- Coronary artery involvement
- Large aneurysms

Available procedures include:

Surgical Procedures

- Bypass grafting
- Endarterectomy
- Aortic reconstruction

Endovascular Procedures

- Balloon angioplasty
- Stent placement
- Stent-graft implantation

Studies indicate that procedures performed during inactive disease have better long-term outcomes and lower restenosis rates.[9],[10]

Pregnancy and Anesthetic Considerations

Pregnancy in women with TA requires careful multidisciplinary management because severe vascular lesions may compromise maternal and fetal circulation.

Both regional and general anesthesia have been used successfully during cesarean section. Continuous monitoring of cerebral perfusion and blood pressure is essential, particularly in patients with significant carotid artery involvement.[11]

Prognosis

Advances in diagnosis, immunosuppressive therapy, biologic agents, and vascular interventions have substantially improved outcomes.

Recent studies report:

- Five-year survival rates exceeding 90%
- Improved vascular outcomes
- Reduced mortality
- Better quality of life

Nevertheless, relapses remain common, particularly among pediatric patients and those with active disease at diagnosis.[3],[6],[9]

Long-term complications include:

- Stroke
- Heart failure

- Renal failure
- Aortic regurgitation
- Aneurysm rupture

Regular follow-up is therefore mandatory.[7]

VI. CONCLUSION

Takayasu arteritis is a rare but potentially devastating large-vessel vasculitis affecting predominantly young women and children. Delayed diagnosis remains a major challenge because early symptoms are nonspecific and laboratory markers may not accurately reflect disease activity. Modern imaging modalities have significantly improved diagnostic accuracy and disease monitoring. Corticosteroids remain the foundation of treatment, while conventional immunosuppressive agents and biologic therapies offer improved disease control. Surgical and endovascular interventions are valuable options for patients with advanced vascular complications. Continued research into disease mechanisms, biomarkers, and targeted therapies is essential to improve outcomes and reduce long-term morbidity associated with this complex disease.

REFERENCES

1. Numano F. The story of Takayasu arteritis. Rheumatology. 2002;41(1):103-106.
2. Russo RAG, Katsicas MM. Takayasu Arteritis. Front Pediatr. 2018;6:265.
3. Keser G, Aksu K, Direskeneli H. Takayasu arteritis: an update. Turk J Med Sci. 2018;48(4):681-697.
4. Parra JR, Perler BA. Takayasu's Disease. Semin Vasc Surg. 2003;16(3):200-208.
5. O'Connor TE, Carpenter HE, Bidari S, Waters MF, Hedna VS. Role of inflammatory markers in Takayasu arteritis disease monitoring. BMC Neurol. 2014;14:62.
6. Aeschlimann FA, Eng SWM, Sheikh S, Laxer RM, Hebert D, Noone D, et al. Childhood Takayasu arteritis: disease course and response to therapy. Arthritis Res Ther. 2017;19:255.
7. Ishikawa K. Natural history and classification of occlusive thromboaortopathy (Takayasu's disease). Circulation. 1978;57(1):27-35.
8. Li Cavoli G, Mulè G, Vallone MG, Caputo F. Takayasu's disease effects on the kidneys: current perspectives. Int J Nephrol Renovasc Dis. 2018;11:225-233.
9. Fields CE, Bower TC, Cooper LT, Hoskin T, Noel AA, Panneton JM, et al. Takayasu's arteritis: operative results and influence of disease activity. J Vasc Surg. 2006;43(1):64-71.
10. Lagneau P, Michel JB, Vuong PN. Surgical treatment of Takayasu's disease. Ann Surg. 1987;205(2):157-166.
11. Clark AG, Al-Qatari M. Anaesthesia for Caesarean section in Takayasu's disease. Can J Anaesth. 1998;45(4):377-379.