BMJ Best Practice

Takayasu's arteritis

The right clinical information, right where it's needed

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Summary

- A vasculitis of large vessels that particularly affects the aorta and its primary branches.
- More common in women and typically presents before the age of 40.
- Typical symptoms include limb claudication on exertion, chest pain, and systemic symptoms of weight loss, fatigue, low-grade fever, and myalgia.
- On examination, vascular bruits may be audible over the carotids, abdominal aorta, or subclavian vessels. Unequal blood pressures may be recorded between sides, and a murmur of aortic regurgitation may be heard if there is aortic root dilation.
- The diagnosis is usually made by vascular imaging.
- Glucocorticoids form the mainstay of treatment, with the additional use of steroid-sparing immunosuppressive agents for resistant disease. Surgery may be required for established complications.
- Long-term complications are due mainly to arterial occlusion and related damage, including limb ischaemia and renal failure.

BASICS

Definition

Takayasu's arteritis is a chronic granulomatous vasculitis affecting large arteries: primarily the aorta and its main branches. Vascular inflammation can cause stenosis, occlusion, and aneurysm formation. Symptoms from vascular ischaemia include claudication and stroke. Diminished or absent pulses and hypertension are common. Constitutional symptoms, including fever and weight loss, are often accompanied by elevation of acute phase markers.[1] [2]

Epidemiology

Takayasu's arteritis is a rare disease. Its distribution is worldwide, although most cases are reported in Asian populations. The reported incidence in the US is 2.6 cases per million population per year in Olmsted County, Minnesota, and in Sweden the incidence has been estimated to be 1.2 cases per million population per year.[5] [9] These figures are likely to underestimate the true prevalence of the disease. Autopsy studies in Japan suggest a higher incidence, with evidence of Takayasu's arteritis in 1 in every 3000 autopsies.[10]

Although Takayasu's arteritis is often thought of as a disease of young women, the disease has variable gender predilection, with women in Japan affected about 8 times more frequently than men, whereas in India men and women are equally represented.[4] The peak incidence is usually in the third decade of life, although among Japanese people it typically presents between the ages of 15 and 25. In European people the mean age at diagnosis is 41.[2]

Disease expression varies in different populations. Compared with Japanese patients, patients in the US are more likely to have constitutional (43% in the US versus 27% in Japan) and musculoskeletal symptoms (53% in the US versus 6% in Japan), claudication (90% in the US versus 13% in Japan), and visual changes (30% in the US versus 6% in Japan).[2]

Aetiology

The aetiology of Takayasu's arteritis is unknown. Environmental and genetic factors are thought to play roles in the development of the disease. Cell-mediated immune mechanisms have been implicated.[1] Genetic screening has shown polymorphisms in IL-12, IL-6, and IL-2 genes in a population of Turkish patients with Takayasu's arteritis.[11] HLA-Bw5 and HLA-B39.2 are reportedly increased in frequency in some populations.[12] [13]

Pathophysiology

Takayasu's arteritis is an immune-mediated vasculitis characterised by granulomatous inflammation of large arteries. Cell-mediated immune mechanisms have been implicated.[1] IL-6 is thought to play an important role in the pathogenesis of Takayasu's arteritis. A small number of patients have been treated with an IL-6 inhibitor with favourable responses.[14]

The immunological and inflammatory response seen in arteries is similar to that observed in large arteries in giant cell arteritis.[1] During the acute phase of vasculitis, inflammation begins in the vasa vasora of the adventitia of muscular arteries.[1] [7] T cells are prominent in the initial cellular response, and anti-endothelial cell antibodies may also be involved.[1] [15] [16]
[Fig-1]

Classification

2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides[3]

Categorises vasculitis based upon the predominant type of vessels involved, and other features including aetiology, pathogenesis, type of inflammation, favoured organ distribution, clinical manifestations, genetic predispositions, and distinctive demographic characteristics.

- Large vessel vasculitis
    - Takayasu's arteritis
    - Giant cell arteritis
- Medium vessel vasculitis
    - Polyarteritis nodosa
    - Kawasaki disease
- Small vessel vasculitis
    - ANCA-associated vasculitis
    - Microscopic polyangiitis
    - Granulomatosis with polyangiitis (Wegener's)
    - Eosinophilic granulomatosis with polyangiitis (Churg-Strauss)
    - Immune complex vasculitis
    - Anti-glomerular basement membrane (anti-GBM) disease
    - Cryoglobulinaemic vasculitis
    - IgA vasculitis (Henoch-Schönlein)
    - Hypocomplementaemic urticarial vasculitis
- Variable vessel vasculitis
    - Behçet's disease
    - Cogan's syndrome
- Single-organ vasculitis
- Vasculitis associated with systemic disease
- Vasculitis associated with probable aetiology

Angiographic classification of Takayasu's arteritis[4]

Classification is based on the vessels involved in the inflammatory process as seen on angiography.

- Type I: Branches of the aortic arch
- Type IIa: Ascending aorta, aortic arch, and branches of the aortic arch
- Type IIb: Ascending aorta, aortic arch, and its branches and thoracic descending aorta
- Type III: Thoracic descending aorta, abdominal aorta, and/or renal arteries
- Type IV: Abdominal aorta and/or renal arteries
- Type V: Features of types IIb and IV

Secondary prevention

As the precise aetiology of Takayasu's arteritis and causes of flare-ups in disease activity are unknown, there are no known specific preventative actions. Management of hypertension is important to prevent further vascular damage. Attention to osteoporosis screening and management is crucial, given the need for glucocorticoid therapy. Patients require influenza and pneumococcal immunisations annually. Use of prophylactic antibiotic therapy to prevent *Pneumocystis jirovecii* pneumonia is important, especially when the prednisolone (prednisone) dose is more than 20 mg daily. Atherosclerotic vascular disease can further complicate the vascular damage caused by Takayasu's arteritis; thus, control of other risk factors is important.

Case history

Case history #1

A 28-year-old woman presents with new left-arm pain. She was previously well but for 2 months has had episodes of low-grade fever, night sweats, and arthralgia. She works as a shop assistant and has noticed left-arm pain when she stocks shelves. Her only medication is an oral contraceptive. She does not smoke cigarettes. On examination, her blood pressure is 126/72 in her right arm, but it cannot be measured in her left arm. The left radial pulse cannot be detected. There is a bruit over the left subclavian artery. Carotid pulses are normal but there is a bruit over the right carotid artery. Femoral and pedal pulses are normal and no abdominal bruits are heard. The left hand is cool but has no other evidence of ischaemia.

Case history #2

A 39-year-old woman presents with headaches of insidious onset over 3 months. She has lost 3 kilograms during this time but feels otherwise well. On examination, bilateral blood pressures taken in the arms are 190/110 on the right and 200/110 on the left. She is taking a multivitamin but no other medications. For the past 20 years she has smoked 10 cigarettes a day. Urinalysis reveals estimated protein of 360 mg/24 hour.

Other presentations

Non-specific constitutional symptoms, including fever, weight loss, and fatigue, are common.[1] [5] Patients may also present with an absent pulse or immeasurable blood pressure in 1 extremity.[6] New-onset hypertension or aortic regurgitation may be present. Coronary artery involvement can lead to angina pectoris, but pericarditis and congestive heart failure are uncommon presentations. Pulmonary artery involvement may result in chest pain, dyspnoea, or haemoptysis. Involvement of cranial arteries can present as a headache, transient ischaemic attack, or stroke. Visual symptoms may include blurring, scotoma, diplopia, and amaurosis fugax. The retinal arteriovenous anastomoses described by Takayasu are rare.[7] Mesenteric artery involvement can cause abdominal pain or gastrointestinal haemorrhage. Vascular bruits are often found on auscultation.[5] [6] Erythema nodosum is occasionally noted.[8]

Step-by-step diagnostic approach

Establishing the diagnosis of Takayasu's arteritis can be difficult, as it may present with non-specific systemic symptoms including fever, night sweats, and weight loss. Other presenting features may include ischaemic symptoms of extremity claudication, transient ischaemic attack, stroke, or chest pain. Laboratory tests are non-specific, reflecting inflammation. Biopsy of involved vessels is not usually feasible, and the diagnosis relies on vascular imaging.[1] [2] [7]

History

In the early stages, constitutional symptoms may include weight loss, low-grade fever, and general fatigue, and these are often ascribed to another cause. The diagnosis of Takayasu's arteritis should be considered in patients under age 40 with symptoms of vascular ischaemia. Although relatively uncommon at presentation, claudication in the upper or lower limb may develop over time. Development of collateral

circulation patterns can lead to a variety of other findings, especially subclavian steal syndrome caused by a stenotic lesion proximal to the origin of the vertebral artery causing lightheadedness on exercise of the upper limb.

Less common manifestations of Takayasu's arteritis include myalgia and arthralgia. Pulmonary arteries are often involved in Takayasu's arteritis but rarely cause symptoms. Chest pain, shortness of breath, and haemoptysis may be due to pulmonary artery stenosis, coronary artery involvement, or heart failure from aortic dilation. Pericarditis is possible but uncommon. Involvement of carotid and vertebral arteries can cause cerebral ischaemia, which may in turn cause visual symptoms (e.g., diplopia, amaurosis fugax, or scotoma), vertigo, lightheadedness, syncope, or headache. History of a previous TIA, or a TIA as a presenting symptom in a young patient, may indicate inflammation of the vertebral or carotid vessels. Less commonly, the mesenteric vessels may be involved, causing abdominal pain and diarrhoea.[1] [2] [7] [17] [18]

Examination

Careful examination of the vascular system is vital. Cool extremities and absent pulses (most commonly the radial pulse) may be noted, and a difference in blood pressure of >10 mmHg on each arm may be significant. If Takayasu's arteritis is suspected, it is advisable to measure the blood pressure on both arms and legs to look for a difference. Hypertension may be a feature, but the blood pressure can also be unusually low if there is a stenosis just proximal to the area of blood pressure measurement. The carotid pulses may feel weaker, and a bruit might be audible. Bruits may also be heard over the supraclavicular region or abdominal aorta. A cardiac murmur may be audible if there is aortic root involvement and aortic regurgitation. Evidence of a previous stroke may suggest CNS involvement. The arteriovenous anastomoses seen on examination of the retina and originally described by Takayasu in 1908 are rarely seen today.[1] [2] [7] Less common manifestations include the appearance of erythema nodosum or pyoderma gangrenosum on the arms or legs.

Investigations

Acute phase markers, including erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP), are usually elevated in patients with active disease. They can be followed as markers of disease activity.[1] [2] [7] Other non-specific markers of inflammation such as normocytic anaemia and thrombocytosis may also be noted. There are no specific laboratory tests for Takayasu's arteritis.

Imaging studies, including computerised tomography, magnetic resonance vascular imaging, and conventional angiography, are currently the most important modalities for establishing a diagnosis of Takayasu's arteritis and can be helpful in monitoring disease activity.
[Fig-2]

[Fig-3]

[Fig-4]

[Fig-5]

Positron emission tomography with radiolabelled fluorodeoxyglucose (PET-FDG) can be used to identify inflammation in the large arteries, and is therefore a useful technique to establish diagnosis. However, the performance of PET-FDG for assessing disease activity over time is still unclear.[19]

Non-invasive vascular ultrasonography is a useful tool for initial evaluation of a patient with suspected Takayasu's arteritis.[1] [2] [7] [18] [20] [21]

Histopathology

Biopsy cannot usually be obtained from one of the vessels typically involved, due to the vessels' size and function as large arteries, and is therefore not part of the usual approach to diagnosis. Biopsy specimens from patients who have undergone revascularisation procedures secondary to complications of Takayasu's arteritis may show histopathological findings identical to those of giant cell arteritis. Temporal artery biopsy is not helpful in making the diagnosis.[1] [2] [7] [18]

Risk factors

Strong

genetic predisposition

- Takayasu's arteritis is most prevalent in Japan, Southeast Asia, India, and Mexico.[1] [2] [9] [10] Polymorphisms in interleukin genes have been demonstrated in a population of Turkish patients with Takayasu's arteritis, and certain HLA antigens have been found in greater-than-expected frequency in some patient populations.[11] [12]

female sex

- Women are affected more often than men in a ratio of approximately 8:1.[1]

age <40

- Patients are usually under age 40 at presentation.[1] [5] [9]

Asian ethnicity

- There are more reported cases in Asian populations compared with non-Asian, although the reasons for this are unclear, and there may be a genetic basis confounding this association.

History & examination factors

Key diagnostic factors

presence of risk factors (common)

- Takayasu's arteritis is more common in people of Asian descent, is 8 times more common in women than in men, and typically presents below age 40.

upper or lower limb claudication (common)

- Progressive symptoms of claudication are more common than claudication as a presenting feature. A history of pain on exertion of the upper or lower limb may be given; upper limb claudication is more common.

absent pulse(s) (common)

- Often found to have unilateral absence of brachial, radial, or carotid pulse due to occlusive disease.

unequal blood pressures (common)

- A discrepancy of >10 mmHg between the 2 arms may be noted.

vascular bruits (common)

- Bruits may be heard over subclavian, carotid, or abdominal vessels due to eccentric flow.

low-grade fever (common)

- A systemic sign often present during the acute phase of inflammation.

Other diagnostic factors

transient ischaemic attack (TIA) (common)

- History of a previous TIA, or a TIA as a presenting complaint in a young patient, may indicate inflammation of the vertebral or carotid vessels.

myalgia (common)

- A systemic symptom, seen in the acute phase; may be accompanied by a rise in inflammatory markers.

arthralgia (common)

- A systemic symptom, seen in the acute phase; may be accompanied by a rise in inflammatory markers.

weight loss (common)

- A systemic symptom, seen in the acute phase; may be accompanied by a rise in inflammatory markers.

fatigue (common)

- A systemic symptom, seen in the acute phase; may be accompanied by a rise in inflammatory markers.

dizziness on upper-limb exertion (common)

- May result from subclavian steal syndrome due to a stenotic lesion developing proximal to the origin of the vertebral artery.

hypertension (common)

- May develop due to involvement and narrowing of the renal arteries. Can be falsely low if there is a narrowed segment proximally.

stroke (uncommon)

- Evidence of a previous stroke may be suggestive of CNS involvement and arteritis affecting the vertebral or carotid vessels.

chest pain (uncommon)

- This can be a feature of Takayasu's arteritis if the coronary vasculature is involved, causing coronary ischaemia, or if pulmonary arteries are affected.

abdominal pain (uncommon)

- May result from involvement of the mesenteric branches of the aorta.

diarrhoea (uncommon)

- May result from involvement of the mesenteric branches of the aorta.

shortness of breath (uncommon)

- Due to pulmonary artery stenosis, coronary artery involvement, or aortic regurgitation.

haemoptysis (uncommon)

- May result from pulmonary artery stenosis or heart failure due to aortic root dilation and aortic regurgitation.

night sweats (uncommon)

- Constitutional symptom that may be reported during the acute phase.

vertigo (uncommon)

- May result from cerebral ischaemia.

syncope (uncommon)

- May result from cerebral ischaemia.

headache (uncommon)

- May result from cerebral ischaemia or hypertension.

heart murmur (uncommon)

- Aortic regurgitation may result from aortic root dilation.

visual symptoms (uncommon)

- Due to cerebral ischaemia secondary to carotid and vertebral involvement. May include amaurosis fugax, scotoma, or diplopia. The retinopathy originally described by Mikoto Takayasu is rarely seen and is usually a late sign.

erythema nodosum (uncommon)

- Uncommonly found on the arms or legs.

pyoderma gangrenosum (uncommon)

- Uncommonly found on the legs.

Diagnostic tests

1st test to order

Test	Result
ESR • A marker of inflammation, but it lacks specificity and sensitivity. Most patients with elevated ESR do not have Takayasu's arteritis, and patients with active disease can have a normal ESR.[1] [2] [18] [22]	**typically >50 mm/hour with active disease**

Test	Result
CRP • A marker of inflammation. Lacks specificity, as it can be raised by any inflammatory process. Sensitivity is moderate.[1] [2] [18] [22]	**typically elevated with active disease**
computerised tomography angiography (CTA) • CT may be performed with helical scanning and 3D reconstruction. It has high sensitivity and specificity (over 95%) for the diagnosis of Takayasu's arteritis. It is preferable to catheter angiogram due to reduced contrast load.[21] [23] [Fig-3] [Fig-4] [Fig-5]	**segmental narrowing or occlusion, occasionally dilation, of affected vessels; aortic aneurysms may be seen; thickening of vessel walls may be seen but is of uncertain significance**
magnetic resonance imaging angiography (MRA) • MR angiography is used to identify arterial involvement, and it may be useful in the assessment of disease activity, with vessel wall thickening and oedema thought to reflect active disease.[24] [Fig-2]	**segmental narrowing, occlusion, or dilation of involved arteries; vessel wall inflammation may be detectable**

Other tests to consider

Test	Result
catheter angiogram • Conventional angiogram using contrast can reveal abnormalities in the aorta and its branches. [Fig-6]	**segmental narrowing or occlusion, occasionally dilation of affected vessels, aortic aneurysms may be seen**
Doppler ultrasound • Particularly useful in the early vascular evaluation of patients with suspected Takayasu's arteritis, as it is a non-invasive procedure. Abdominal ultrasound may reveal mesenteric or renal artery stenosis, and transthoracic/trans-oesophageal studies can detect abnormalities in the upper aorta and subclavian and carotid arteries.[25]	**segmental narrowing, occlusion, and/or dilation of involved arteries**
positron emission tomography with radiolabelled fluorodeoxyglucose (PET-FDG) • Can be used to identify inflammation in the large arteries and is therefore a useful technique to establish diagnosis. However, the performance of PET-FDG for assessing disease activity over time is still unclear.[19] • PET tracer uptake has been correlated with areas of active disease on MR angiography and has been correlated with elevation of ESR/CRP. The sensitivity and specificity of PET imaging has not been established; atherosclerotic lesions may also show increased tracer uptake.[21] [26] [27]	**may show increased uptake in actively inflamed arterial segments**

Differential diagnosis

Condition	Differentiating signs / symptoms	Differentiating tests
Giant cell arteritis (GCA)	• Patients are usually older; average age is 74 years. May have polymyalgic syndrome with proximal myalgia. Jaw claudication is common. Lower extremity involvement is less common.	• Imaging with CT or MR angiography; GCA is more likely to have cranial artery involvement and less likely to have lower extremity involvement.
Essential hypertension	• Intact pulses and the absence of bruits. No marked difference in blood pressure between each side.	• Clinical diagnosis. • No stenoses on vascular imaging.
Syphilis	• Firm, painless ulcer at site of primary inoculation, usually genital region. Symmetrical non-itchy rash accompanies systemic symptoms.	• Positive syphilis serology. • Catheter or CT angiogram: typical calcification of the proximal ascending aorta.
Tuberculosis (TB)	• Persistent productive cough. Recent travel to an endemic area.	• Mantoux test: elicits delayed hypersensitivity reaction, which will be positive in people with latent infection, previously cleared infection, or in those previously immunised. • CXR: may show evidence of pulmonary TB foci. • Sputum culture: takes 4 to 12 weeks to culture acid-fast bacilli but is diagnostic if found. • QuantiFERON® gold test: blood test to detect interferon gamma production when incubated with TB bacilli. Can identify active and latent infection.
Spondyloarthropathy	• Back pain and stiffness lasting more than 1 hour, particularly in the mornings. Peripheral arthritis. May be preceded by urethritis or cervicitis in the case of reactive arthritis. Accompanying symptoms may include psoriasis, palmar-plantar pustulosis, iritis, uveitis, or conjunctivitis.	• X-ray of spine: may demonstrate sacroiliitis. • X-ray of peripheral joint affected by arthritis may show periarticular osteolysis in psoriatic arthritis.

Condition	Differentiating signs / symptoms	Differentiating tests
Behçet's disease	• A triad of oral and genital ulceration with uveitis. Often accompanied by a peripheral arthritis. May have thrombotic arterial and venous occlusions.	• Angiography: reveals saccular dilation of involved arteries or thrombotic occlusion. • CSF examination supportive but not diagnostic; increased inflammatory cells and protein.
Kawasaki disease	• Typically affects children under age 5. High-grade fever with strawberry-tongue-marked lymphadenopathy. Red eyes with uveitis or conjunctivitis. Rash and peeling of the skin on the palms and soles may be seen.	• Clinical diagnosis using set criteria. • Angiography reveals saccular dilation of coronary arteries if affected.
Marfan's syndrome	• Typically tall people with long limbs. May have a family history of Marfan's syndrome. Susceptible to lens dislocation. • Systemic signs and symptoms absent.	• Clinical diagnosis. • Family history. • Genetic testing rarely carried out.
Ehlers-Danlos syndrome	• May have hypermobile joints or paper-thin skin scars. • Systemic signs and symptoms absent.	• Angiography: if vascular wall collagen affected, may reveal saccular dilation of involved arteries. • Genetic testing.
Atherosclerosis	• More common in men, may have associated risk factors of hypertension, smoking, diabetes, and raised cholesterol. Typically, patients are over age 40.	• Angiography: typical abrupt narrowing of artery rather than tapered narrowing. Lesions usually at the vessel origin and carotid bifurcations.
Fibromuscular dysplasia	• Pulses present but may be diminished. Hypertension common and most commonly affects renal and carotid arteries.	• Angiography: characteristic beading of affected arteries. Aorta not usually involved.

Diagnostic criteria

American College of Rheumatology 1990 criteria for the classification of Takayasu's arteritis[22]

These are not formal diagnostic criteria but were established to help differentiate Takayasu's arteritis from other forms of vasculitis. Imaging may include conventional angiography or MR or CT angiography. Presence of 3 or more criteria has a sensitivity of 90.5% and a specificity of 97.8% for diagnosis of Takayasu's arteritis.

- Age at onset of disease less than or equal to 40 years.
- Claudication of extremities: development and worsening of fatigue and discomfort in muscles of 1 or more extremity while in use.
- Decreased brachial artery pulse.
- Systolic BP difference greater than 10 mmHg between arms.
- Bruit over subclavian arteries or abdominal aorta.
- Arteriographic abnormality: narrowing or occlusion of the entire aorta, its primary branches or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or similar causes; changes usually focal or segmental.

K. Ishikawa: proposed criteria for the clinical diagnosis of Takayasu's arteriopathy[28]

Criteria suggested by Ishikawa for diagnosing Takayasu's arteritis were based on observations in 108 Japanese patients. In addition to the presence of the obligatory criterion, the presence of 2 major, 4 minor, or 1 major plus 2 minor criteria suggests a high probability of Takayasu's disease with 84% sensitivity. These criteria are not widely applied.

Obligatory criterion:

- Age less than or equal to 40 years

Major criteria:

- Lesion of the left mid subclavian artery
- Lesion of the right mid subclavian artery

Minor criteria:

- High ESR
- Common carotid artery tenderness
- Hypertension
- Aortic regurgitation or annulo-aortic ectasia
- Lesions of the pulmonary artery
- Lesions of the left mid common carotid artery
- Lesions of the distal brachiocephalic trunk
- Lesions of the thoracic aorta
- Lesions of the abdominal aorta.

Step-by-step treatment approach

The goal of treatment for Takayasu's arteritis is to manage systemic symptoms and suppress vascular inflammation to prevent damage to vessels and the tissues they supply. Glucocorticoids are the mainstay of treatment, with immunosuppressive agents used for resistant patients or those with glucocorticoid-related side effects. Surgical or percutaneous revascularisation procedures may be required to improve blood flow or prevent rupture of aneurysms. Low-dose aspirin should be considered to help prevent ischaemic complications.

Initial presentation

Oral prednisolone (prednisone) should be started. No studies have established the optimal way to taper prednisolone (prednisone), but most follow the regimen originally described in the cohort from the National Institutes of Health.[6] [29] A common tapering regimen is to reduce prednisolone (prednisone) by 5 mg/week until reaching a dose of 20 mg/day. Thereafter, the taper rate is decreased to 2.5 mg/week until reaching a dose of 10 mg/day. Thereafter, the dose is lowered by 1 mg/day each week, as long as disease does not become more active.[6] Pulse intravenous glucocorticoids have been tried in some patients with CNS symptoms, but there are no data to support their use.

During treatment, patients should be evaluated regularly by clinical examination and measurement of inflammatory markers (i.e., erythrocyte sedimentation rate and C-reactive protein); initially, this may be every few days. Vascular imaging studies such as CT or MR angiography should be performed every 3 to 12 months during the active phase of treatment and annually thereafter. Low-dose aspirin should be considered to help prevent ischaemic complications.

A critical issue is in trying to determine whether or not disease is active. Constitutional symptoms are frequent when Takayasu's arteritis is active. However, lack of such symptoms does not mean that the disease is inactive. New vascular lesions can develop even when no other signs, symptoms, or laboratory features of disease activity are present. If new vascular lesions are seen, the disease is considered to be active. Conversely, acute phase markers may be elevated from other causes and not indicate disease activity. Careful evaluation is required.

Refractory to, or unable to wean from, glucocorticoids

The majority of patients will go into remission with glucocorticoid therapy. However, relapse occurs in more than 50% of patients during dose tapering. Immunosuppressive or cytotoxic therapy is used for patients with Takayasu's arteritis who are unresponsive to glucocorticoid therapy or who relapse with dose tapering. This can be with or without concomitant glucocorticoid therapy. Agents such as methotrexate, azathioprine, and mycophenolate can be used.[30] In patients with severe refractory disease, use of cyclophosphamide may be indicated.[31]

Refractory to glucocorticoids and additional immunosuppression

Tumour necrosis factor (TNF)-alpha inhibitors, primarily infliximab, have also been used successfully in the management of Takayasu's arteritis when immunosuppressive agents and glucocorticoid therapy have failed to control disease activity. These agents are used in addition to glucocorticoids as a steroid-sparing agent.[32] [33] [34] Non-biological immunosuppressive agents may be continued or tapered.[35]

Symptoms of intermittent claudication or ischaemic organ dysfunction

Surgical intervention may be required in patients with severe complications of Takayasu's arteritis. Vascular lesions are usually not reversible with immunosuppression alone. Therefore, patients with significant limb claudication or severe ischaemic organ dysfunction may require surgical intervention. Percutaneous angioplasty can be effective in the short term, but re-stenosis is common.[36] Good long-term outcomes have been reported with vascular bypass surgery.[37] [38] Patients with progressive dilation of the aorta may require surgical repair.[39]

Prevention of complications from long-term glucocorticoid use

Prolonged glucocorticoid treatment can lead to significant morbidity. Attention to potential side effects is critical. Long-term glucocorticoid therapy increases the risk of osteoporosis, and the greatest amount of bone loss occurs in the first 6 to 12 months of therapy. Prevention of glucocorticoid-induced bone loss by treatment with calcium, vitamin D, and bisphosphonates is recommended. Glucocorticoid-induced diabetes mellitus is a potential side effect of therapy, and a high index of suspicion is required.

Due to the immunosuppressive effects of glucocorticoids, influenza and pneumococcal vaccination are recommended, and, while the daily prednisolone (prednisone) dose is greater than 20 mg/day, prophylaxis for *Pneumocystis jirovecii* pneumonia with trimethoprim/sulfamethoxazole is advised.

Treatment details overview

Consult your local pharmaceutical database for comprehensive drug information including contraindications, drug interactions, and alternative dosing. (see Disclaimer)

Acute (summary)		
Patient group	**Tx line**	**Treatment**
all patients	**1st**	**glucocorticoids**
	plus	**low-dose aspirin**
	plus	**bone protection therapy**
	adjunct	**immunosuppressants**
	adjunct	**pneumocystis pneumonia prophylaxis**
	adjunct	**evaluation for surgery or endovascular procedure**
with persistent active disease	**plus**	**tumour necrosis factor (TNF)-alpha antagonist**

Treatment options

Acute

Patient group	Tx line	Treatment
all patients	**1st**	**glucocorticoids**

» Glucocorticoids are the mainstay of therapy to suppress vascular inflammation and systemic inflammatory symptoms. Duration of therapy varies, but dose can be reduced once signs and symptoms have diminished and acute phase markers have normalised.

» A common tapering regimen is to reduce prednisolone (prednisone) by 5 mg/week until reaching a dose of 20 mg/day. Thereafter, the taper rate is decreased to 2.5 mg/week until reaching a dose of 10 mg/day. Thereafter, the dose is lowered by 1 mg/day each week, as long as disease does not become more active.[6]

» Glucocorticoid tapering may have to be stopped and the dose increased if there is return of disease activity.

» Specialist consultation is recommended for guidance on paediatric dosing.

Primary options

» prednisolone: 1 mg/kg/day orally initially, then taper according to response

plus **low-dose aspirin**

» Low-dose aspirin is recommended to reduce the risk of organ damage from vascular ischaemia. It is usually stopped 1 week prior to any surgical procedure.

Primary options

» aspirin: 75 mg orally once daily

plus **bone protection therapy**

» Long-term glucocorticoid therapy increases the risk of osteoporosis, and the greatest amount of bone loss occurs in the first 6 to 12 months of therapy. Risk is proportional to cumulative glucocorticoid dose, so dose should be reduced as soon as possible. It is important to prevent bone loss by ensuring adequate dietary calcium intake and prophylactic use of bisphosphonates and vitamin D3/calcium supplementation.

Acute

Patient group	Tx line	Treatment
		» Specialist consultation is recommended for guidance on paediatric dosing. **Primary options** » alendronic acid: 5 mg orally once daily **OR** **Primary options** » alendronic acid: 35 mg orally once weekly **--AND--** » calcitriol: 0.25 micrograms orally once daily **--AND--** » calcium carbonate: 1000-1500 mg/day orally given in 2-3 divided doses
	adjunct	**immunosuppressants** » Relapse occurs in more than 50% of patients during glucocorticoid tapering. Therefore, adjunctive use of immunosuppressive agents such as methotrexate, azathioprine, or mycophenolate is often necessary. » In an open-label study, methotrexate was effective as a steroid-sparing agent for a subset of patients with Takayasu's arteritis.[30] » Azathioprine or mycophenolate can be considered for patients who are intolerant to, or relapse while on, methotrexate.[6] » In patients with severe, refractory, and life-threatening disease, use of cyclophosphamide may be indicated.[31] » There is no evidence to advise on discontinuation of immunosuppressive therapy, and this will depend on individual circumstances. » Specialist consultation is recommended for guidance on paediatric dosing. **Primary options** » methotrexate: 15-25 mg orally/ subcutaneously once weekly on the same day each week **--AND--** » folinic acid: 10 mg orally every 6 hours for 10 doses starting 10 hours after methotrexate dose **-or-** » folic acid: 1 mg orally once daily

Acute

Patient group	Tx line	Treatment
		OR
		Secondary options
		» azathioprine: 2 mg/kg/day orally
		OR
		Secondary options
		» mycophenolate mofetil: 1 to 1.5 g orally/ intravenously twice daily
		OR
		Tertiary options
		» cyclophosphamide: 2 mg/kg/day orally
	adjunct	**pneumocystis pneumonia prophylaxis**
		» While patients are receiving over 20 mg/day of prednisolone (prednisone), prophylaxis for pneumocystis pneumonia is recommended. Trimethoprim/sulfamethoxazole is the recommended antibiotic.
		» Some recommend continuing prophylaxis, as long as the CD4 lymphocyte count is <200 cells/ mm^3.
		» Specialist consultation is recommended for guidance on paediatric dosing.
		Primary options
		» trimethoprim/sulfamethoxazole: 160/800 mg orally three times weekly
	adjunct	**evaluation for surgery or endovascular procedure**
		» Vascular lesions are not usually reversible with immunosuppression alone. Therefore, patients with significant limb claudication or severe ischaemic organ dysfunction may require surgical intervention. Some reports have indicated good short-term outcomes with percutaneous intervention for vascular stenoses in patients with Takayasu's arteritis. However, re-stenoses are common with this intervention, and bypass surgery often yields better long-term results.[6] [36]
		» Patients with aneurismal disease of the aorta may require surgical repair.[39] Patients with active disease requiring operation are more likely to require revision.[37] Therefore, control of inflammation prior to vascular surgery is

Acute

Patient group	Tx line	Treatment
		important to improve surgical outcome. Long-term complications may include anastomotic aneurysms and congestive heart failure.[39]
with persistent active disease	**plus**	**tumour necrosis factor (TNF)-alpha antagonist** » TNF-alpha antagonist therapy may be considered with persistent active disease despite treatment with glucocorticoids and immunosuppressive agents. » In a pilot study of relapsing Takayasu's arteritis, anti-TNF therapy resulted in improvement in 93% of patients and sustained remission in 67%. Most patients in this study were treated with infliximab.[32] » Retrospective reviews also support the use of anti-TNF therapy for refractory disease.[33] [34] » There is no evidence to advise on discontinuation of immunosuppressive therapy, and this will depend on individual circumstances. » Specialist consultation is recommended for guidance on paediatric dosing. **Primary options** » infliximab: 3-5 mg/kg intravenously as a single dose at 0, 2, and 6 weeks, then every 4-8 weeks thereafter

Emerging

Interleukin-6 (IL-6) blockade

IL-6 is thought to play an important role in the pathogenesis of Takayasu's arteritis. Several small case series of patients treated with an IL-6 inhibitor (tocilizumab) have been reported. In general, most patients have had favourable responses, although disease relapse may still occur.[14] [35] Further investigation is required.

Abatacept

T-cell-mediated mechanisms are involved in the pathogenesis of Takayasu's arteritis. Abatacept inhibits activation of T cells. However, in one randomised clinical trial, addition of abatacept to a treatment regimen with prednisolone (prednisone) did not reduce the risk of relapse in patients with Takayasu's arteritis.[40]

Recommendations

Monitoring

The monitoring interval should vary inversely with the level of disease activity, being shorter for those with more active disease. However, as the disease may become active without new constitutional symptoms, regular follow-up is needed. In addition to history and physical examination, ESR, CRP, and FBC should be checked at each visit. Vascular imaging studies such as CT or MR angiography should be performed every 3 to 12 months during the active phase of treatment and annually thereafter.

Patient instructions

Measures to help with control of hypertension, such as following a low-salt diet, are important to prevent damage to the arteries leading to stroke, heart attack, or kidney failure. A programme of gradually increasing exercise can help form a collateral circulation, which provides new pathways for blood to reach organs and limbs and lessens claudication symptoms. Stopping smoking and controlling blood fats, including cholesterol, is essential to good general health and to the health of the arteries. General health measures also include keeping immunisations up to date, especially if the patient is maintained on immunosuppressive therapy. [Vasculitis Foundation] [Medline Plus: Takayasu arteritis]

Complications

Complications	Timeframe	Likelihood
peripheral vascular ischaemia	**variable**	**high**
Many of the complications of Takayasu's arteritis represent ischaemic symptoms related to the development of vascular stenoses and occlusions. Differentiating between ischaemia resulting from active vasculitis and ischaemia from vascular damage can be difficult. Regular vascular imaging studies can help with follow-up but should also be obtained in the setting of new ischaemic symptoms. Attempts to control vascular inflammation are needed to try to minimise long-term vascular damage.		
hypertension	**variable**	**high**
Hypertension is a common complication, usually due to renal artery or aortic valve stenosis.[44]		
osteoporosis secondary to glucocorticoid use	**variable**	**high**
Long-term glucocorticoid therapy increases the risk of osteoporosis, and the greatest amount of bone loss occurs in the first 6 to 12 months of therapy. Risk is proportional to cumulative dose, so glucocorticoid dose should be reduced as soon as possible.		
diabetes mellitus secondary to glucocorticoid use	**variable**	**medium**
Long-term glucocorticoid therapy can cause the development of diabetes. A high degree of vigilance is required.		
Pneumocystis jirovecii pneumonia	**variable**	**medium**

FOLLOW UP

Complications	Timeframe	Likelihood
Patients require influenza and pneumococcal immunisations annually. Use of prophylactic antibiotic therapy to prevent *Pneumocystis jirovecii* pneumonia is important, especially when the prednisolone (prednisone) dose is more than 20 mg daily.		
aortic aneurysm	**variable**	**medium**
Most often involves the ascending thoracic aorta.		
aortic regurgitation	**variable**	**medium**
Aortic valve insufficiency, usually due to aortic root dilation, is found in about 25% of patients.[44]		
congestive heart failure	**variable**	**medium**
Congestive heart failure occurs in about 25% of patients.[44]		
angina	**variable**	**low**
Angina from coronary artery involvement is described in up to 10% of patients.[44]		
stroke	**variable**	**low**
Involvement of carotid or vertebral arteries can result in a TIA or stroke. Visual disturbance including blurred vision and amaurosis fugax may be present, but permanent visual loss is uncommon.[44]		

Prognosis

Remission of disease is usually defined as the lack of clinical and laboratory features of disease, with no evidence of new vascular lesions on follow-up imaging examinations.[2] [6] Most patients achieve disease remission, although the majority require immunosuppressive therapy in addition to glucocorticoids.[6] Monophasic disease is described in about 20% of patients.[2] In one series, sustained remission, lasting for at least 6 months while on <10 mg of prednisolone (prednisone) daily, was attained by only 28% of patients, and only 17% remained in remission after prednisolone (prednisone) was discontinued.[6]

Disease relapses occur in >80% of all patients who go into remission.[2] [6] Relapses can occur despite ongoing immunosuppressive treatment. Relapses manifest as new vascular lesions on imaging studies are typically associated with elevation of acute phase markers, but this laboratory evidence of active disease can be lacking.[6] [41]

Mortality and morbidity

Cardiac failure is a common cause of death.[18] Long-term morbidity is related primarily to complications from vascular ischaemia. Symptomatic extremity claudication occurs in about 50% of patients. Upper-extremity claudication is more common than lower-extremity symptoms. Thoracic aortic aneurysm, aortic valve involvement, and arteritis of coronary and pulmonary arteries are known complications that are associated with increased mortality. The 5-year mortality in Takayasu's arteritis is estimated to be between 70% and 93%.[42]

Pregnancy

Because Takayasu's arteritis is primarily a disease of young women, pregnancy is often a consideration. There are few data about pregnancy in patients with Takayasu's arteritis, but successful pregnancies have been reported.[6] [43] In one series of patients, the annual incidence of pregnancies fell after the diagnosis of Takayasu's arteritis, and the percentage of miscarriages showed an upward trend.[43] Careful management of hypertension is necessary during pregnancy.

Diagnostic guidelines

North America

Criteria for the classification of Takayasu arteritis

Published by: American College of Rheumatology **Last published:** 1990

Summary: There are no established guidelines for the diagnosis of Takayasu's arteritis. These criteria serve to differentiate Takayasu's arteritis from other forms of vasculitis. Presence of 3 or more criteria has a sensitivity of 90.5% and a specificity of 97.8% for diagnosis of Takayasu's arteritis.

Online resources

1. Vasculitis Foundation *(external link)*

2. Medline Plus: Takayasu arteritis *(external link)*

REFERENCES

Key articles

- Weyand CM, Goronzy JJ. Medium- and large-vessel vasculitis. N Engl J Med. 2003 Jul 10;349(2):160-9. Abstract
- Kerr GS. Takayasu's arteritis. Rheum Dis Clin North Am. 1995 Nov;21(4):1041-58. Abstract
- Hall S, Barr W, Lie JT, et al. Takayasu arteritis. A study of 32 North American patients. Medicine (Baltimore). 1985 Mar;64(2):89-99. Abstract
- Kissin EY, Merkel PA. Diagnostic imaging in Takayasu arteritis. Curr Opin Rheumatol. 2004 Jan;16(1):31-7. Abstract
- Arend WP, Michel BA, Block DA, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum. 1990 Aug;33(8):1129-34. Full text Abstract

References

1. Weyand CM, Goronzy JJ. Medium- and large-vessel vasculitis. N Engl J Med. 2003 Jul 10;349(2):160-9. Abstract
2. Kerr GS. Takayasu's arteritis. Rheum Dis Clin North Am. 1995 Nov;21(4):1041-58. Abstract
3. Jennette JC, Falk RJ, Bacon PA, et al. 2012 revised International Chapel Hill Consensus Conference nomenclature of vasculitides. Arthritis Rheum. 2013 Jan;65(1):1-11. Full text Abstract
4. Moriwaki R, Noda M, Yajima M, et al. Clinical manifestations of Takayasu arteritis in India and Japan - new classification of angiographic findings. Angiology. 1997 May;48(5):369-79. Abstract
5. Hall S, Barr W, Lie JT, et al. Takayasu arteritis. A study of 32 North American patients. Medicine (Baltimore). 1985 Mar;64(2):89-99. Abstract
6. Maksimowicz-McKinnon K, Clark T, Hoffman GS. Limitations of therapy and a guarded prognosis in an American cohort of Takayasu arteritis patients. Arthritis Rheum. 2007 Mar;56(3):1000-9. Full text Abstract
7. Numano F, Okawara M, Inomata H, et al. Takayasu's arteritis. Lancet. 2000 Sep 16;356(9234):1023-5. Abstract
8. Werfel T, Kuipers JG, Zeidler H, et al. Cutaneous manifestations of Takayasu arteritis. Acta Derm Venereol. 1996 Nov;76(6):496-7. Abstract
9. Waern AU, Anderson P, Hemmingsson A. Takayasu's arteritis: a hospital-region based study on occurrence, treatment and prognosis. Angiology. 1983 May;34(5):311-20. Abstract

10. Nasu T. Takayasu's truncoarteritis in Japan. A statistical observation of 76 autopsy cases. Pathol Microbiol (Basel). 1975;43(2-O):140-6. Abstract

11. Saruhan-Direskeneli G, Biçakçigil M, Yilmaz V, et al. Interleukin (IL)-12, IL-2, and IL-6 gene polymorphisms in Takayasu's arteritis from Turkey. Hum Immunol. 2006 Sep;67(9):735-40. Abstract

12. Kimura A, Kitamura H, Date Y, et al. Comprehensive analysis of HLA genes in Takayasu arteritis in Japan. Int J Cardiol. 1996 Aug;54 Suppl:S61-9. Abstract

13. Sahin N, Aksu K, Kamali S, et al. PTPN22 gene polymorphism in Takayasu's arteritis. Rheumatology (Oxford). 2008 May;47(5):634-5. Abstract

14. Clifford A, Hoffman GS. Recent advances in the medical management of Takayasu arteritis: an update on use of biologic therapies. Curr Opin Rheumatol. 2014 Jan;26(1):7-15. Abstract

15. Seko Y, Sato O, Takagi A, et al. Restricted usage of T-cell receptor Valpha-Vbeta genes in infiltrating cells in aortic tissue of patients with Takayasu's arteritis. Circulation. 1996 May 15;93(10):1788-90. Full text Abstract

16. Eichhorn J, Sima D, Thiele B, et al. Anti-endothelial cell antibodies in Takayasu arteritis. Circulation. 1996 Nov 15;94(10):2396-401. Full text Abstract

17. Yoneda S, Nukada T, Tada K, et al. Subclavian steal in Takayasu's arteritis. A hemodynamic study by means of ultrasonic Doppler flowmetry. Stroke. 1977 Mar-Apr;8(2):264-8. Abstract

18. Mwipatayi BP, Jeffery PC, Beningfield SJ, et al. Takayasu arteritis: clinical features and management: report of 272 cases. ANZ J Surg. 2005 Mar;75(3):110-7. Abstract

19. Treglia GM. Usefulness of whole-body fluorine-18-fluorodeoxyglucose positron emission tomography in patients with large-vessel vasculitis: a systematic review. Clin Rheumatol. 2011 Oct;30(10):1265-75. Abstract

20. Webb M, Chambers A, Al-Nahhas A, et al. The role of 18F-FDG PET in characterising disease activity in Takayasu arteritis. Eur J Nucl Med Mol Imaging. 2004 May;31(5):627-34. Abstract

21. Kissin EY, Merkel PA. Diagnostic imaging in Takayasu arteritis. Curr Opin Rheumatol. 2004 Jan;16(1):31-7. Abstract

22. Arend WP, Michel BA, Block DA, et al. The American College of Rheumatology 1990 criteria for the classification of Takayasu arteritis. Arthritis Rheum. 1990 Aug;33(8):1129-34. Full text Abstract

23. Yamada I, Nakagawa T, Himeno Y, et al. Takayasu arteritis: evaluation of the thoracic aorta with CT angiography. Radiology. 1998 Oct;209(1):103-9. Abstract

24. Aluquin VP, Albano SA, Chan F, et al. Magnetic resonance imaging in the diagnosis and follow up of Takayasu's arteritis in children. Ann Rheum Dis. 2002 Jun;61(6):526-9. Full text Abstract

25. Andrews J, Al-Nahhas A, Pennell DJ, et al. Non-invasive imaging in the diagnosis and management of Takayasu's arteritis. Ann Rheum Dis. 2004 Aug;63(8):995-1000. Full text Abstract

REFERENCES

26. Meller J, Strutz F, Siefker U, et al. Early diagnosis and follow-up of aortitis with [(18)F]FDG PET and MRI. Eur J Nucl Med Mol Imaging. 2003 May;30(5):730-6. Abstract

27. Zerizer I, Tan K, Khan S, et al. Role of FDG-PET and PET/CT in the diagnosis and management of vasculitis. Eur J Radiol. 2010 Mar;73(3):504-9. Abstract

28. Ishikawa K. Diagnostic approach and proposed criteria for the clinical diagnosis of Takayasu's arteriopathy. J Am Coll Cardiol. 1988 Oct;12(4):964-72. Abstract

29. Kerr GS, Hallahan CW, Giordano J, et al. Takayasu arteritis. Ann Intern Med. 1994 Jun 1;120(11):919-29. Abstract

30. Hoffman GS, Leavitt RY, Kerr GS, et al. Treatment of glucocorticoid-resistant or relapsing Takayasu arteritis with methotrexate. Arthritis Rheum. 1994 Apr;37(4):578-82. Abstract

31. Shelhamer JH, Volkman DJ, Parrillo JE, et al. Takayasu's arteritis and its therapy. Ann Intern Med. 1985 Jul;103(1):121-6. Abstract

32. Hoffman GS, Merkel PA, Brasington RD, et al. Anti-tumor necrosis factor therapy in patients with difficult to treat Takayasu arteritis. Arthritis Rheum. 2004 Jul;50(7):2296-304. Full text Abstract

33. Molloy ES, Langford CA, Clark CE, et al. Anti-tumor necrosis factor therapy in patients with refractory Takayasu arteritis: long-term follow-up. Ann Rheum Dis. 2008 Nov;67(11):1567-9. Abstract

34. Schmidt J, Kermani TA, Bacani AK, et al. Tumor necrosis factor inhibitors in patients with Takayasu arteritis: Experience from a referral center with long-term follow-up. Arthritis Care Res (Hoboken). 2012 Jul;64(7):1079-83. Abstract

35. Koster MJ, Matteson EL, Warrington KJ. Recent advances in the clinical management of giant cell arteritis and Takayasu arteritis. Curr Opin Rheumatol. 2016 May;28(3):211-7. Abstract

36. Rao SA, Mandalam KR, Rao VR, et al. Takayasu arteritis: initial and long-term follow-up in 16 patients after percutaneous transluminal angioplasty of the descending thoracic and abdominal aorta. Radiology. 1993 Oct;189(1):173-9. Abstract

37. Fields CE, Bower TC, Cooper LT, et al. Takayasu's arteritis: operative results and influence of disease activity. J Vasc Surg. 2006 Jan;43(1):64-71. Full text Abstract

38. Labarca C, Makol A, Crowson CS, et al. Retrospective comparison of open versus endovascular procedures for takayasu arteritis. J Rheumatol. 2016 Feb;43(2):427-32. Abstract

39. Miyata T, Sato O, Koyama H, et al. Long-term survival after surgical treatment of patients with Takayasu's arteritis. Circulation. 2003 Sep 23;108(12):1474-80. Full text Abstract

40. Langford CA, Cuthbertson D, Ytterberg SR, et al; Vasculitis Clinical Research Consortium. A randomized, double-blind trial of abatacept (CTLA-4Ig) for the treatment of Takayasu arteritis. Arthritis Rheumatol. 2017 Apr;69(4):846-53. Abstract

41. Maksimowicz-McKinnon K, Hoffman GS. Takayasu arteritis: what is the long-term prognosis? Rheum Dis Clin North Am. 2007 Nov;33(4):777-86. Abstract

42. Phillip R, Luqmani R. Mortality in systemic vasculitis: a systematic review. Clin Exp Rheumatol. 2008 Sep-Oct;26(5 Suppl 51):S94-104. Abstract

43. Vanoli M, Daina E, Salvarani C, et al; Itaka Study Group. Takayasu's arteritis: a study of 104 Italian patients. Arthritis Rheum. 2005 Feb 15;53(1):100-7. Full text Abstract

44. Maksimowicz-McKinnon K, Clark T, Hoffman GS. Limitations of therapy and a guarded prognosis in an American cohort of Takayasu arteritis patients. Arthritis Rheum. 2007;56:1000-1009. Full text Abstract

Images

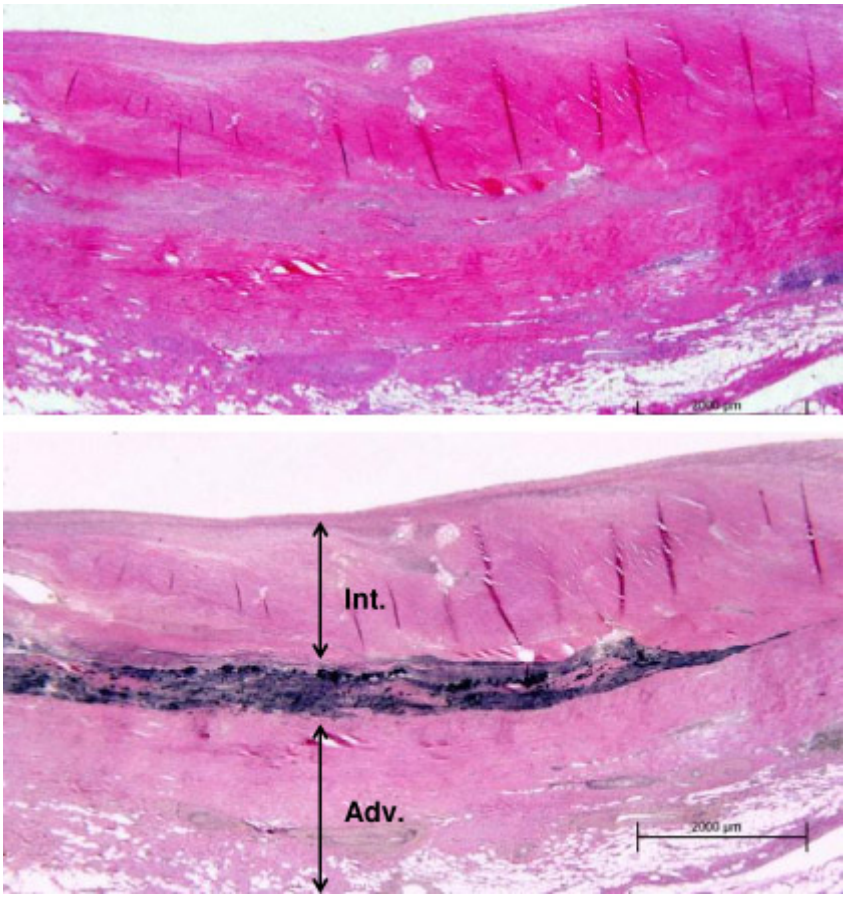

Figure 1: Photomicrograph of the aorta from a patient with Takayasu's arteritis demonstrates marked thickening of the intimal layer and inflammatory infiltrates in the media and laminar necrosis

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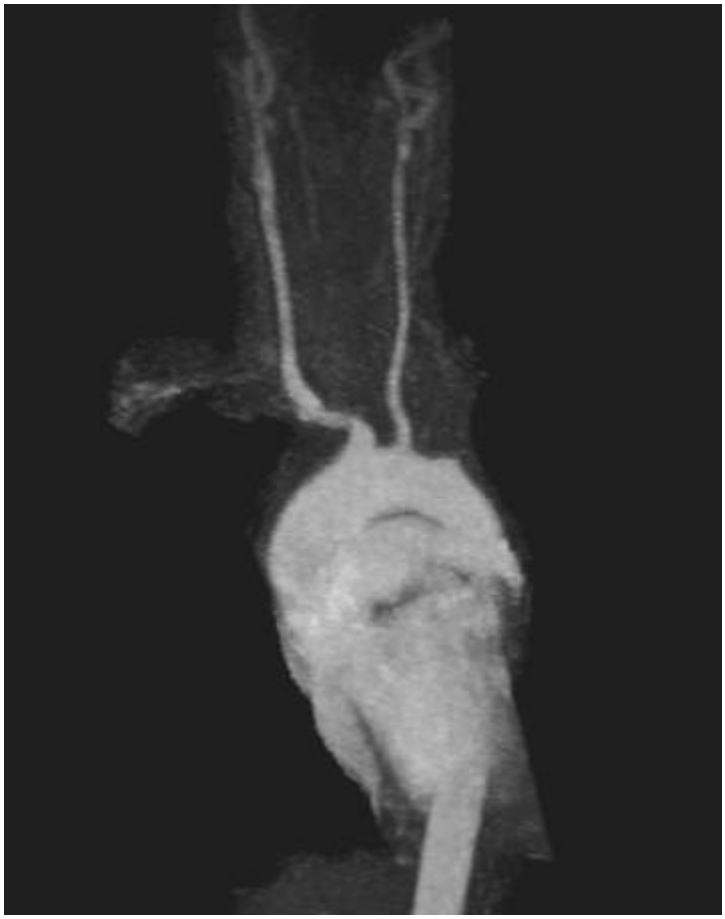

Figure 2: MR angiogram of the aortic arch and major vessels showing occlusion of bilateral subclavian arteries; left common carotid artery has small diameter; proximal vertebral arteries are not identified

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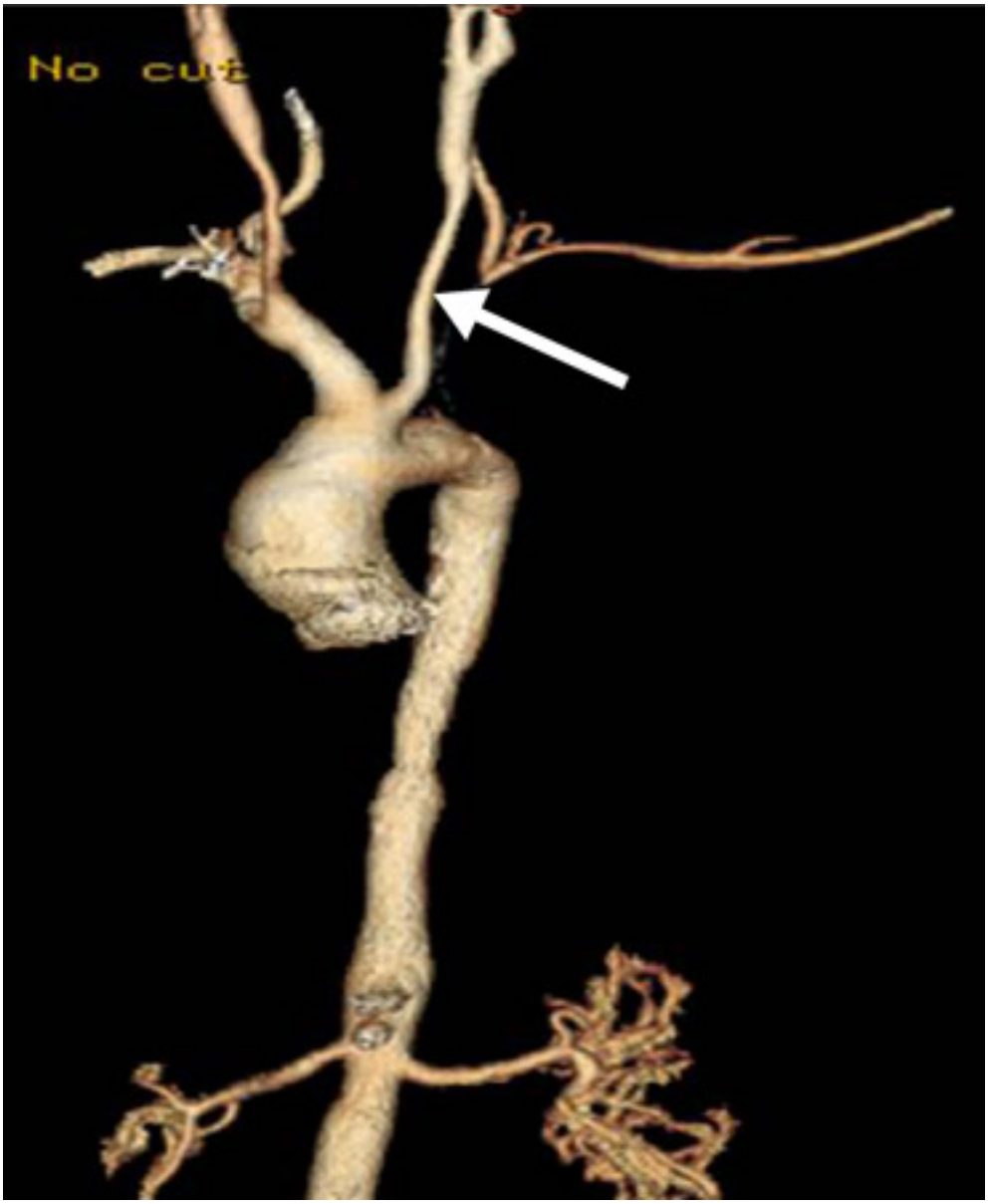

Figure 3: CT angiogram, with 3D reconstruction of the aortic arch and major vessels, showing proximal occlusion of the left subclavian artery and patent left vertebral artery distal to the occlusion (left vertebral steal syndrome)

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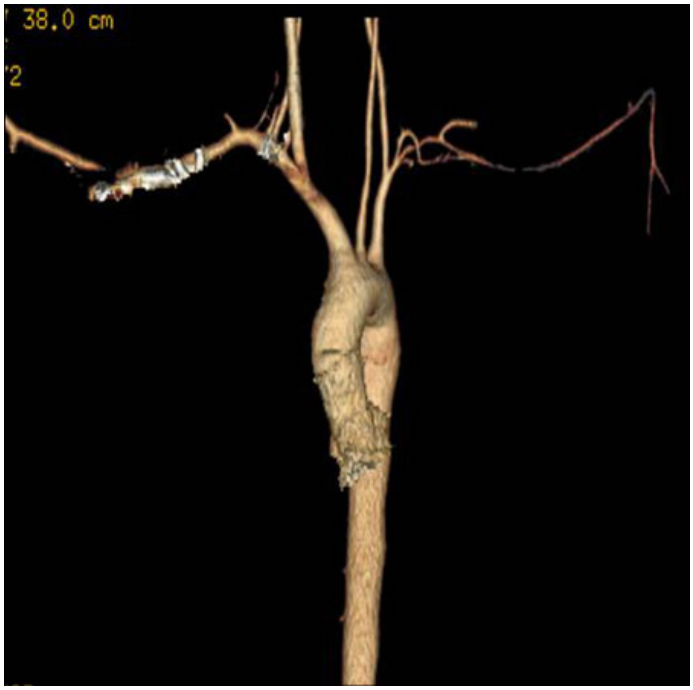

Figure 4: CT angiogram, with 3D reconstruction of the aortic arch and major vessels, showing narrowing of the left common carotid artery and left subclavian artery

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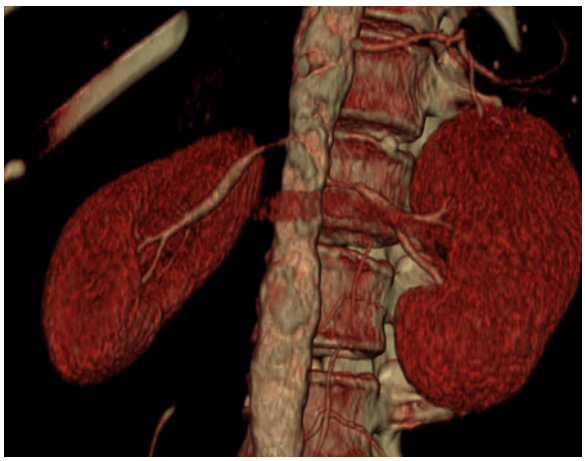

Figure 5: CT angiogram with 3D reconstruction showing bilateral renal artery stenosis

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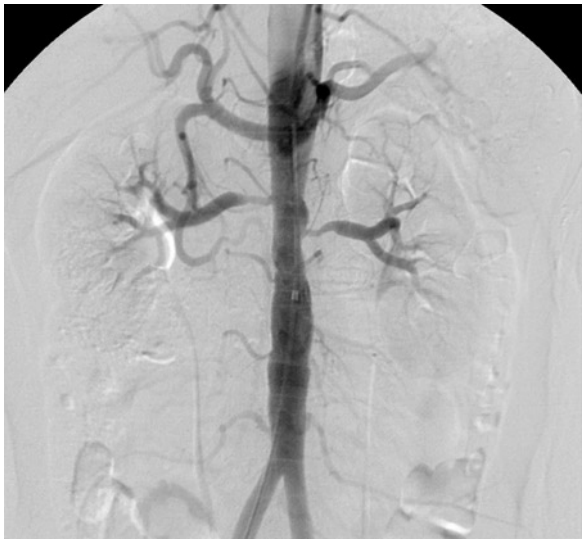

Figure 6: Catheter angiogram showing bilateral renal artery stenosis

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