

Takayasu's arteritis: a review of the literature and the role of endovascular treatment



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KEYWORDS

- abdominal aortic aneurysm stent/prosthesis
- aneurysm
- renal artery stenosis
- thoracic aorta aneurysm

Abstract

Takayasu's arteritis (TA) is a chronic non-specific vasculitis with variable presentation in different ethnicities and countries. Treatment options vary and are dependent on the stage and presentation of the disease. We aimed to review current literature related to TA, focusing on the role of endovascular treatment in revascularisation. The temporal course of the disease and stage at presentation influence the management of TA. Treatment options include medical therapy, endovascular intervention or surgical vascular reconstruction. The decision to intervene is individualised according to vascular anatomy and the presence of haemodynamically significant lesions. There are currently no clear guidelines regarding the choice between the endovascular and open surgical approaches, but studies have shown that endovascular procedures are associated with slightly higher rates of restenosis while surgical procedures have higher rates of thrombosis. Peri-procedural immunosuppression is suggested if the disease is active at the point of intervention. This improves outcomes but at the cost of immunosuppression-related side effects. Careful long-term follow-up is essential due to the risk of disease activation or flare-up, requiring appropriate evaluation of the diseased vessels.

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Abbreviations

CABG	coronary artery bypass graft
CRP	C-reactive protein
CTA	computed tomography angiography
MACE	major adverse cardiac events
MRA	magnetic resonance angiography
PCI	percutaneous coronary intervention
PET	positron emission tomography
PTA	percutaneous transluminal angioplasty
TA	Takayasu's arteritis
TARAS	Takayasu's arteritis-induced renal artery stenosis

Introduction

Takayasu's arteritis (TA) is a chronic non-specific granulomatous large-vessel vasculitis, which affects the aorta and its main branches. It may also affect the coronary and pulmonary arteries^{1,2}. TA has historically been described as having a strong association with female patients, though the degree of association with the female gender may vary for different populations³. In the Japanese population where TA was originally described, the majority (80-90%) of the TA patient population consists of females. By comparison, Indian, Thai and Israeli populations demonstrate greater gender heterogeneity, with a larger (31-38%) proportion of male patients^{3,4}. The disease commonly presents in the second or third decade of life regardless of ethnicity, although a small percentage may present in childhood^{2,4,5}. The incidence of TA has been described worldwide as up to 3.3 per million, and is generally considered to be most common in Asia⁶.

TA has been described as the infiltration of inflammatory cells into the adventitia and media resulting in a cell-mediated immune response, involving NK T cells and CD4 T cells which form characteristic granulomas and giant cells. This is different from atherosclerotic lesions which consist of the accumulation of lipid-laden foam cells involving the intima layer. These features make atherosclerotic lesions respond better to percutaneous transluminal angioplasty (PTA) as compared with TA lesions, with 15.5% residual stenosis after subclavian artery PTA in TA compared with only 8.3% in atherosclerotic lesions⁷.

CLINICAL MANIFESTATIONS

In a series of 60 patients described by Kerr et al, only 33% had systemic symptoms on presentation⁵. Hypertension is most often associated with renal artery stenosis. Sixty-eight percent (68%) of patients had extensive vascular disease; stenotic lesions were 3.6-fold more common than were aneurysms (98% compared with 27%)⁵. Most of the aneurysms were located in the thoracic aorta (83.3%), with a smaller proportion (19%) in the abdominal aorta⁸. It has been noted that the site of affected lesions differs amongst various countries, therefore causing different sets of symptoms. Occlusive disease seems to be more prevalent in Japan, the USA, and Europe, whereas aneurysmal disease is more common in India, Thailand, Mexico, and Africa⁹.

The clinical course of TA can be subdivided into two phases - the early or active phase, and the late, chronic, or inactive phase¹⁰. During the early or active phase, the initial intimal inflammation process is followed by oedema and subsequent infiltration of lipids and blood cells. The resultant calcification and intimal thickening that follows the inflammatory phase represents the chronic phase of TA, which is similar to atherosclerosis. Clinically, constitutional symptoms are predominant during the acute phase, whereas the chronic phase is characterised by symptoms related to arterial compromise¹¹. Though TA patients have often been described as having high rates of relapse despite being on medical treatment¹², a small cohort study (n=26) demonstrated that a significant proportion of patients (76%) had no angiographically significant relapse over more than three years¹³.

Geographically, TA demonstrated significant anatomical variability in terms of the site of the disease. This can potentially be explained by the heterogeneous genetic presentation of TA patients around the globe¹⁴. In Japan and South America, cervical and thoracic arterial lesions are more prevalent, but in Israel and other Asian countries abdominal lesions are more frequent¹⁵. In a study of 106 Japanese patients, the presentation was as follows: 41.5% had thoracic aorta lesions, 31.1% had abdominal aorta lesions, 22.6% had moderate to severe aortic regurgitation, 21.7% renal artery lesions, 8.5% had coronary lesions, 4.7% pulmonary artery lesions and 2.8% had loss of vision¹⁶. Japanese patients often present with "pulseless disease", with the majority of the stenotic lesions occurring in the ascending aorta, the aortic arch, and/or its branches (58%) and occasionally extending into the thoracic and abdominal aorta^{4,17}. On the other hand, in Korean patients, vasculitis generally occurs in the abdominal aorta (30%) involving renal arteries^{4,17,18}. Indian patients tend to present with hypertension, with a number of studies having noted the characteristic involvement of the abdominal aorta and the renal arteries in the majority of cases (71-92%)¹⁹⁻²¹. In an Indian observational study, patients with early TA were observed to have isolated abdominal and renal artery lesions at presentation, which over the course of 10-20 years progressed to involve the entire aorta and its branches⁴. Approximately 20-26% of Indian patients with TA have aneurysmal lesions of the abdominal aorta²², with the only case series of 30 patients with the aneurysmal form of TA being published in India in 1990²³. In the UK, a study of 97 patients revealed that the majority of patients (95%) had arterial stenosis or occlusions, as compared to the other 5% who had a discrete aneurysm²⁴. The supra-aortic vessels were involved more frequently, comprising 45.9% of affected segments, with the next most common site being the aorta (25.3%)²⁴. Examples of anatomic involvement on imaging scans are illustrated in **Figure 1-Figure 4**.

PRESENTATION

In Japanese patients, more common presentations include aortic regurgitation, ischaemic heart disease and visual disturbances, while amongst Korean and Indian patients the more frequent

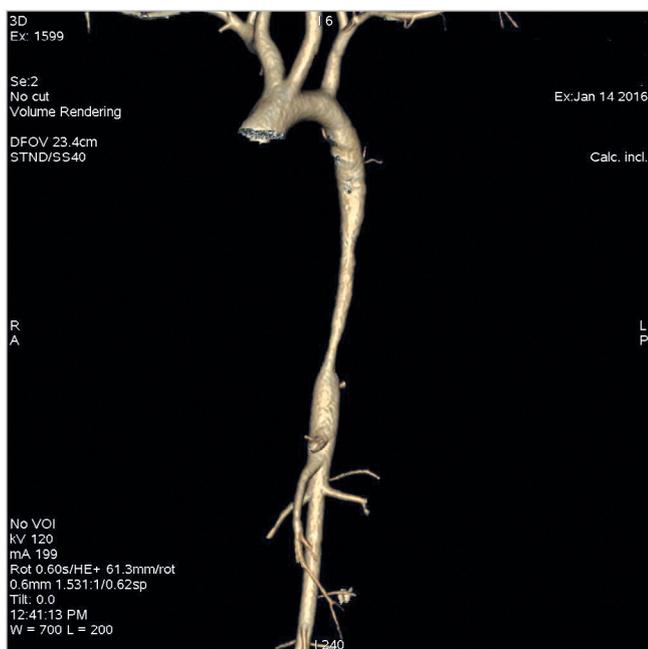


Figure 1. A 7-year-old female with stenosis of the descending thoracic aorta demonstrated on a CT aortogram.

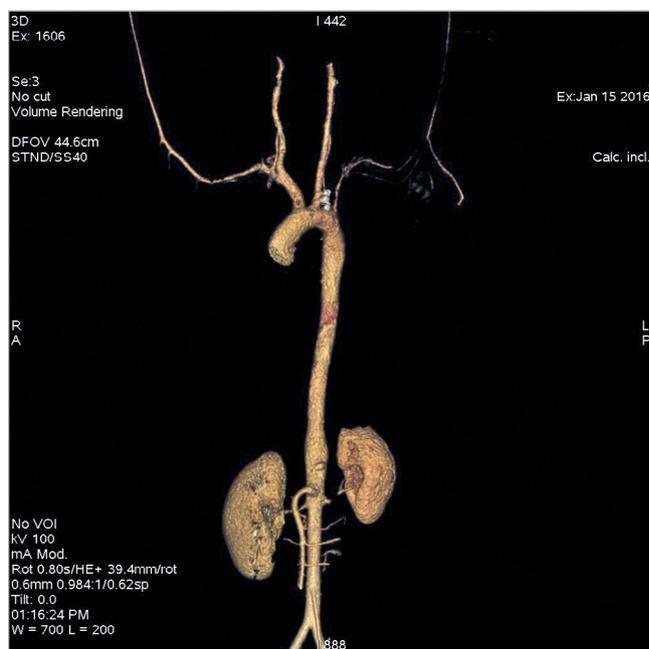


Figure 2. CT aortogram of a 12-year-old male showing stenosis of the left subclavian artery and left renal artery.

presentations were hypertension, headache, exertional dyspnoea, dizziness and malaise^{4,17,18}. In a study of 106 patients in India, hypertension was the most common mode of presentation (51.3%) and was detected in 82 patients (77.4%) at the time of presentation. Other presentations include vascular bruits which were heard in 72 patients (67.9%), while 13 (12.3%) patients were found to be in congestive heart failure²⁰. In Beijing, 530 patients were studied,

of whom 60% had hypertension, 57.5% had vascular bruits in the upper abdomen, 47.4% had carotid bruits, 37.2% had pulse deficit at the extremities and 24.7% had intermittent claudication²⁵. In essence, the affected lesions vary in different countries and therefore present differently.

The differences in clinical presentation based on geographical distribution are summarised in **Table 1**.

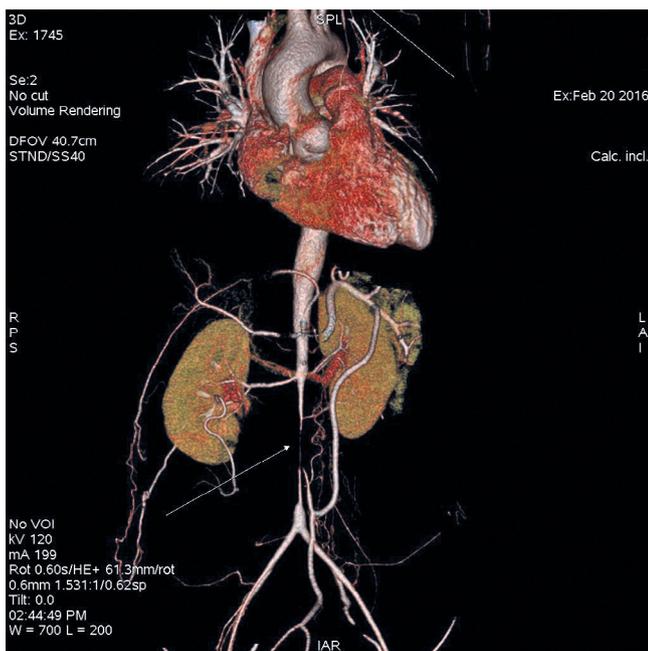


Figure 3. A 23-year-old female with stenosis of the descending abdominal aorta with collaterals from celiac to inferior mesenteric artery.



Figure 4. A CT aortogram showing bilateral common carotid artery aneurysms.

Table 1. Common presentation of Takayasu's arteritis.

Country	Symptoms	Site of affected lesions
Korea ¹⁷	<ul style="list-style-type: none"> - Headache (60%) - Exertional dyspnoea (42%) - Dizziness (36%) - Malaise or weakness (34%) - Hypertension 	<ul style="list-style-type: none"> - Abdominal aorta (46%) - Descending thoracic aorta (37%) - Ascending aorta (1%) - Aortic arch (2%)
Japan ^{16,23}	<ul style="list-style-type: none"> - Aortic regurgitation (22.6%) - Ischaemic heart disease (8.5%) - Visual disturbances/loss (2.8%) 	<ul style="list-style-type: none"> - Thoracic aorta (41.5%) - Abdominal aorta (31.1%) - Moderate to severe aortic regurgitation (22.6%) - Renal artery lesions (21.7%) - Coronary lesions (8.5%) - Pulmonary artery lesions (4.7%)
India ¹⁹	<ul style="list-style-type: none"> - Hypertension (51.3%) - Vascular bruit (67.9%) - Congestive heart failure (12.3%) 	<ul style="list-style-type: none"> - Type I (branches of aortic arch) (6.6%) - Type II (aortic arch, its branches and descending thoracic aorta) (6.6%) - Type III (descending thoracic aorta and abdominal aorta) (3.8%) - Type IV (abdominal aorta only) (27.3%) - Type V (aortic arch, descending thoracic aorta and abdominal aorta) (55.7%)
China ²⁴	<ul style="list-style-type: none"> - Hypertension (60%) - Vascular bruits upper abdomen (57.5%) - Carotid bruits (47.4%) - Pulse deficit (37.2%) - Intermittent claudication (24.7%) 	<ul style="list-style-type: none"> - Type I (20.8%) - Type II (37.1%) - Type III (42.1%) - Type IV (52.3%)

DIAGNOSIS

The first set of diagnostic criteria for TA was initially established by Ishikawa in 1988. This was then replaced by a new set of criteria by the American College of Rheumatology (ACR) in 1990²⁶. The current ACR classification consists of six criteria: (1) onset at age less than or equal to 40 years, (2) claudication of an extremity, (3) decreased brachial artery pulse, (4) greater than 10 mmHg difference in systolic blood pressure between arms, (5) a bruit over the subclavian arteries or the aorta, and (6) arteriographic evidence of narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities. The presence of at least three of the six criteria was found to have demonstrated a sensitivity of 90.5% and a specificity of 97.8%²⁶.

A modification to the 1988 Ishikawa criteria by Sharma et al was validated and compared to the ACR 1990 criteria in a small Indian population (n=106) with a reported higher sensitivity (96% vs. 77.4%) and similar specificity²⁷. However, this has not been validated in other populations/studies; the ACR criteria remain the most widely used in studies and clinical practice^{14,28-31}.

The current “gold standard” investigation is digital subtraction angiography (DSA); however, it is invasive and only identifies late, structural changes in the vasculature³². Angiographic findings can be classified into six types, according to the vessels involved (**Figure 5**)^{33,34}. Additionally, involvement of the coronary and pulmonary arteries is indicated as C (+) or P (+), respectively. Type V has been documented as the most common type³⁵.

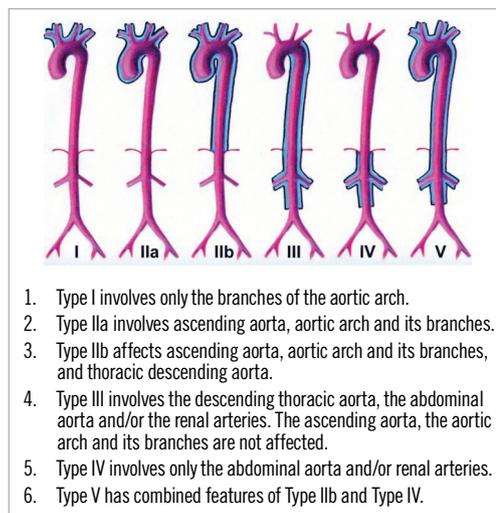


Figure 5. Angiographic types of Takayasu's arteritis.

Recent advances in non-invasive vascular imaging, on the other hand, have provided new insights into TA²⁶. Contrast-enhanced computed tomography angiography and magnetic resonance angiography are useful in demonstrating vascular anatomy, wall enhancement, oedema, as well as thickening, which might enable early disease detection while the luminal diameter is still preserved. 18F-fluorodeoxyglucose positron emission tomography has also been found to provide important additional information by highlighting areas of increased metabolic activity and is therefore useful to detect inflammation with a reported high sensitivity and specificity in TA, allowing diagnosis of early pre-stenotic disease^{36,37}. However, there are limitations with this technique, which include a lack of standardised technique for quantification of uptake, limited availability, and lack of reliable evidence for evaluation of disease activity³⁸. Histological diagnosis of TA includes granulomatous arteritis with infiltration of Langhans giant cells in the media with smooth muscle cell necrosis and destruction of the internal elastic membrane³⁹.

TREATMENT OPTIONS

Current treatment options include medical therapy, endovascular intervention and surgical vascular reconstruction. Briefly, the modality of treatment is largely dependent on the individual patient's clinical course and extent of disease activity. Patients who present during the active stage of the disease require corticosteroids and immunosuppressive agents to curb the systemic and vascular inflammatory response, whereas patients who have either

progressed into or present during the chronic phase of TA may require revascularisation, either by an endovascular or by an open approach, for haemodynamically significant arterial lesions⁴⁰.

MEDICAL TREATMENT

The administration of corticosteroids and other immunosuppressive agents has demonstrated positive anti-inflammatory effects in a majority of patients with TA¹. Although glucocorticoids have long been considered the mainstay of treatment for TA due to excellent initial response rates of 40-93%, sustained remission is maintained in only 28% of cases and up to 80% of patients experience steroid-related adverse effects^{1,5,28,41-43}. Immunosuppressive drugs such as cyclophosphamide, methotrexate and azathioprine are usually added due to glucocorticoid resistance, relapse upon reduction of glucocorticoid dose or serious side effects from steroid therapy¹. About 40% of all steroid-resistant patients respond to the addition of cytotoxic agents²⁸. Although remission can be achieved in the majority of patients with immunosuppression, over 90% have some form of relapse¹².

Biologic agents such as anti-TNF- α agents, anti-IL-6R, anti-CD20, anti-IL-12/23 p40, and the soluble CTLA4 receptor fusion protein have recently been used in patients with resistant TA, including those patients who fail to achieve remission despite glucocorticoid steroids and other immunosuppressive therapy. A recent meta-analysis in 2014, based on three small randomised controlled trials, showed that anti-TNF agents (infliximab, etanercept and adalimumab) are not effective in the ability to induce remission or reduce the amount of corticosteroids required⁴⁴. However, the same meta-analysis also observed that data from numerous case series suggested that tocilizumab and infliximab may be effective in the management of large vessel vasculitis and refractory TA⁴⁴, with 37-67% of patients achieving complete remission and 53% having a partial response⁴⁵⁻⁴⁷. Adverse events such as infections and hypersensitivity in approximately 20% of patients result in discontinuation of anti-TNF- α therapy⁴⁵.

INTERVENTION

TIMING

Intervention in TA is often challenging due to the complexity of the lesions and the high rates of restenosis. Approximately 20% of patients are resistant to any kind of medical treatment^{28,46}. This in turn leads to the need for endovascular or surgical interventions, which are usually recommended at a time of quiescent TA^{10,28,29}. This can be achieved by opting to revascularise patients during the chronic inactive phase or by administering preprocedural immunosuppression, which has been found to lower restenosis rates^{24,28,48,49}. Studies have demonstrated that both endovascular and open surgery during active TA were associated with a higher incidence of arterial complications as well as the need for repeat revascularisation^{10,28,29,50}. Therefore, assessment of disease activity plays an important role in determining the optimal timing for revascularisation. Clinical symptoms coupled with haemoglobin levels, erythrocyte sedimentation rate (ESR)

and C-reactive protein (CRP), as well as radiological imaging (e.g., CT angiography, magnetic resonance angiography, 18F-FDG positron emission tomography), can be used for assessment of disease activity^{28,36,51}.

INDICATIONS AND PATIENT SELECTION

Broadly speaking, the decision to intervene in TA has to be individualised based on the anatomic location of disease of each patient. For each lesion, i.e., renal artery stenosis, subclavian artery stenosis or aortic regurgitation, the decision to intervene is based on standard indications (**Table 2**)². Indications for intervention are generally the presence of haemodynamically significant lesions, such as hypertension caused by severe renal artery stenosis, severe limb claudication, progressive aneurysm enlargement, cerebrovascular ischaemia or critical stenosis of three or more cerebral vessels, coronary artery ischaemia, moderate to severe aortic regurgitation and severe aortic coarctation^{5,16,52}. For abdominal aneurysms, the indications are similar to those due to atherosclerosis and connective tissue disease. There are no other special situations that will alter the indication for surgery². Patients with two or more major complications of TA, defined as Ishikawa's prognostic criteria stage III, were noted to derive the most benefit from revascularisation as compared to those with less extensive disease^{52,53}.

Table 2. Intervention of Takayasu's arteritis based on organ involvement^{5,15,51}.

Anatomy	Intervention	Indication for intervention
Aorta	1) Open surgery 2) Percutaneous endovascular repair	– Progressive aneurysm enlargement – Moderate to severe aortic regurgitation – Severe aortic coarctation
Renal artery stenosis	1) Percutaneous angioplasty 2) Open surgery/reconstruction	– Hypertension
Coronary artery stenosis	1) Percutaneous intervention with drug-eluting stent and immunosuppressive steroids 2) CABG	– Coronary artery ischaemia
Carotid and subclavian artery occlusion	1) Open surgery 2) Percutaneous angioplasty	– Severe limb claudication – Cerebrovascular ischaemia – Critical stenosis of 3 or more cerebral vessels

ENDOVASCULAR VERSUS OPEN APPROACH

Currently there are no clear guidelines regarding the selection of endovascular versus open surgical intervention in TA patients with chronic inactive disease. The decision hinges on many factors including the anatomy of the lesion, local practices and expertise, and perioperative risk.

In general, a number of case control and cohort studies have reported lower restenosis rates for patients who underwent open

surgery as compared to an endovascular approach^{24,28,31,41,54,55}. A review of recent comparative studies published from 2007 to 2015 showed that 15-50% of patients who underwent surgery had recurrent disease requiring revision surgery after 5-20 years. For endovascular interventions, the restenosis rates ranged from 17-70% at 5-10 years. Conversely, a small US cohort study in 2009 found that treatment of late inactive stage TA lesions with either just angioplasty or angioplasty with stenting resulted in excellent to good clinical improvement at follow-up at 46.8 months⁵⁶. Symptom recurrence occurred in 31.4%, which was then successfully treated with repeat angioplasty and stenting⁵⁶. The contrasting, limited and retrospective evidence suggests that there may not be a “one size fits all” approach to revascularisation. Instead, each patient and lesion must be evaluated individually to determine the best mode of revascularisation.

In the modern era, survival rates for patients undergoing either open or endovascular revascularisation are generally good: 94.3% at 6.5 years and 73.5% at 20 years^{10,24,29,50,52}. According to Saadoun, the rates of early complications (<30 days) of surgical and endovascular procedures were similar in terms of restenosis, thrombosis and stroke²⁸. However, in terms of late complications (>30 days), endovascular procedures had slightly higher rates of restenosis at 64.5% compared to 46.1%, while surgical procedures had higher rates of thrombosis (15.4% versus 3.2%)²⁸. For surgically treated patients, it is important to identify a disease-free area of artery for the bypass graft anastomosis, to prevent the development of anastomotic strictures and/or the formation of pseudoaneurysms postoperatively⁵⁶. The high restenosis rates published are similar to those quoted in another recent 25-year retrospective cohort study (open surgery 44%, endovascular approach 66%, $p=0.33$)⁵⁴. This implies that, regardless of the approach, restenosis remains a frequent late complication and may necessitate repeat intervention.

RENAL ARTERY STENOSIS

The optimal approach for the treatment of Takayasu’s arteritis-induced renal artery stenosis (TARAS) remains a topic for discussion, with evidence to support both endovascular and open approaches. Aortorenal bypass was shown in two earlier studies (2003-2004) to have five-year patency rates of 79% with a post-operative morbidity of 19% and 17%, respectively^{57,58}. More recently, primary angioplasty of TARAS has demonstrated procedural success of more than 90%, five-year patency of 67-91% and lower complication rates (5.7% in one study and 0% in another). Stenting is occasionally performed for ostial lesions, long-segment lesions, or incomplete dilatation of stenosis and dissection^{10,59-61}. Given the similar long-term outcomes of both approaches coupled with the reduced complication rate of endovascular techniques, angioplasty is currently favoured by most centres⁵⁰.

CORONARY ARTERY STENOSIS

Coronary stenosis in patients with TA tends to be ostial, involving the left main artery. A recent small retrospective analysis

suggests that percutaneous coronary intervention (PCI) and coronary artery bypass graft (CABG) have similar long-term outcomes in patients with TA and stable disease. Patients with active TA who went for CABG had a lower incidence of major adverse cardiac events (MACE) as compared to PCI⁶². Use of periprocedural immunosuppression for three months prior to PCI has been found to maintain coronary stent patency for up to 10 years²⁴.

CAROTID AND SUBCLAVIAN ARTERY OCCLUSION

Common carotid and subclavian artery lesions in TA are often long, irregular and fibrotic. Surgical bypass has superior patency compared to endovascular intervention but with higher rates of major complications^{24,63}. Endovascular intervention is usually reserved for stenoses less than 5 cm, with initial success rates of up to 81% when combined with periprocedural immunosuppression⁶³⁻⁶⁵.

THORACIC AND ABDOMINAL AORTA

Historically, aneurysmal disease of the aorta in TA was treated with open surgery to varied outcomes⁶⁶⁻⁶⁸. In contemporary practice, it is now commonly treated with standard techniques and devices such as aortic stent grafts^{69,70}.

PROGNOSIS

A literature review revealed the overall survival at 15 years after diagnosis of TA to be 82.9%⁵³. Aortic regurgitation, retinopathy, aneurysm and secondary hypertension are associated with poorer prognosis, the presence of two or more of which led to higher five-year (69.9%) and 10-year (36.7%) mortality rates^{71,72}. Common causes of death include congestive cardiac failure, renal failure, cerebrovascular accident, and pulmonary infections.

Conclusion

Takayasu’s arteritis is a chronic non-specific granulomatous vasculitis affecting the aorta and its main branches. Its presentation is different in different ethnicities and countries. Management includes medical therapy with immunosuppressive therapy during the acute phase. Surgical bypass/reconstruction or endovascular therapy may be required for haemodynamically significant or symptomatic disease during the late phase. Careful long-term follow-up is essential due to the risk of disease activation or flare-up, requiring appropriate evaluation of the diseased vessels⁵⁶.

Impact on daily practice

Endovascular interventions are recommended at a time of quiescent TA, which can be achieved by timing them during the chronic inactive phase or by administering preprocedural immunosuppression. Limited data exist but suggest that surgery results in lower restenosis rates than endovascular intervention. Primary angioplasty for TARAS is currently favoured by most centres, due to procedural success rates of more than 90% and lower complications as compared to the open approach.

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Permission for the use of the image labelled **Figure 5** was obtained from RadioGraphics.

Conflict of interest statement

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