



# Surgical treatment of Takayasu's disease

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## ■ ABSTRACT

Takayasu's disease affects young females in the second and third decade of life. During the chronic phase, the aorta and its major arteries become stenotic, causing significant sequelae. Surgical treatment is possible with expectation of good results. The author advises treatment of stenotic arteries that are potentially dangerous or that adversely affect lifestyle with either percutaneous transluminal angioplasty or surgery. The author emphasizes the difference between surgical procedures for atherosclerosis versus a procedure for Takayasu's disease.

## I. INTRODUCTION

Takayasu's disease (TD) is an unusual illness that affects young females in the second and third decade of life by causing inflammation of the major large arteries.<sup>1</sup> The disease has two phases. The acute phase presents with systemic signs and symptoms of inflammation including fatigue, weight loss, fever, loss of appetite, and pain over involved arteries. This acute phase is infrequently diagnosed and sometimes confused with a viral illness. The disease then progresses to a more chronic phase. During this time stenotic lesions develop in the thoracic and abdominal aorta and its major branches. The patient then presents with signs and symptoms depending on the location of the involved arteries. This chronic stage could cause differences in blood pressure in the upper extremities; bruits over the carotid and subclavian arteries and abdominal aorta; weak distal pulses; angina; myocardial infarction; hypertension; and transient ischemic attacks or strokes.

Although there is a growing experience with magnetic resonance angiography and CT angiography, standard arteriography is still the preferred method of diagnosis.<sup>2-5</sup> Since this disease can affect arteries above and below the diaphragm, it is recommended that all arteriographic studies on patients with the diagnosis of TD include the major arterial system from the arch of the aorta to both femoral arteries. Characteristic findings on arteriography include long, tapered stenoses that show a smooth intimal lining that involves the entire artery and not just the bifurcations. The subclavian, axillary, carotid, and to a lesser extent the abdominal aorta and renal arteries are the arteries most commonly involved.

The decision to recommend surgery can be difficult. These patients are young and frequently less tolerant of lifestyle changes forced upon them by TD. Involvement of a major arterial system makes them susceptible to significant medical sequelae including stroke, hypertension, congestive heart failure, and myocardial infarction. However, the surgery itself is complicated and has its own risks and complications. Obviously the final decision will require careful consultation between the attending physicians and the patient.

## Basic principles of surgical treatment

Takayasu's disease is not atherosclerosis. Therefore surgical treatment is different for patients with TD than for patients with arterial problems associated with atherosclerosis. The following are general principles to be considered in patients with TD who are candidates for surgery:

1. Patients with TD are frequently young. One should not assume that because the patient is young, he or she may not have significant medical problems. The multi-arterial involvement by TD could mean that the individual has significant renal disease, cardiac disease, and other problems that could affect the overall surgical outcome. Therefore, a complete preoperative evaluation is essential.
2. It is important that surgeons be conservative with the management of patients with TD. Long-term prognosis for patients with TD properly treated is actually quite good.<sup>6</sup> Surgery should be used only if there is a very significant problem that would affect a patient's prognosis or significantly interfere with the patient's lifestyle.
3. Percutaneous transluminal angioplasty (PTA) has become an effective alternative to surgery.<sup>6</sup> Initially, there was considerable concern about the long-term overall results of PTA. Dilating a chronic lesion might initially be successful, but long-term follow-up might show a restenosis. Restenosis does occur, but with the development of stents, the long-term incidence of restenosis may be considerably less. Therefore, in the initial management of patients with TD who are candidates for operation, consultation with appropriate radiologic interventionists to determine the feasibility of PTA is appropriate.
4. The basic surgical procedure is a bypass operation. The bypass should always be between arteries uninvolved with TD. Arteries that appear normal on arteriography should be used for the proximal and distal anastomoses. It is believed that the incidence of anastomotic problems in arteries that are without dis-

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ease will be considerably low. However, note that even if the artery looks normal on arteriography, there is still a 44% incidence of microscopic involvement with TD, so these patients could still have anastomotic problems in the future.<sup>7</sup>

5. Endarterectomy is almost never performed on patients with TD. The pathology involves all three layers of the artery, and removing the intima and part of the media is difficult and at times impossible.
6. Emergency or urgent surgery is usually not necessary and should be avoided. In general, patients with TD develop chronic lesions over a long period of time and, therefore, do not have the benefit of collateral formation.
7. Surgery should be avoided during the acute phases of the illness. Most reports suggest but do not prove that anastomotic problems are less likely if the patient's illness is quiescent.

## II. CEREBROVASCULAR DISEASE

The major branches of the aortic arch are the arteries most commonly affected with TD. Patients with atherosclerosis commonly have a stroke due to emboli originating from the carotid bifurcation. However, patients with TD do not have focal narrowings of atherosclerosis but instead have long tapered narrowings that may suddenly occlude, causing a stroke.

To understand this, consider the normal anatomy of the aortic arch. The three main arteries off the arch are the innominate, the left carotid, and the left subclavian. All three vessels provide important blood flow to the brain. The innominate gives off the right carotid and the right vertebral, so essentially all circulation for the right side of the brain comes from the innominate. The left carotid and the left subclavian, which gives off the left vertebral, provides circulation to the left side of the brain. Involvement of these major arteries and their branches is quite common. It is not unusual to have patients in their initial arteriography show occlusion of one or more of these major arteries. Therefore, if one or more arteries are occluded and other arteries are involved, sudden occlusion of the involved arteries could cause a catastrophic loss of blood flow to the brain. In the author's original presentation, four patients presented with cerebrovascular accidents and all four of these patients had occlusion of one or more thoracic aortic arch arteries.<sup>8</sup>

Patients who present with these multiple occlusions of the major thoracic aortic arch arteries present a significant dilemma. They may not be symptomatic. Their arteries may be involved but not occluded. Nevertheless, the author feels that if there is significant narrowing or occlusion of major arteries such as the innominate, left carotid, subclavian, or their branches, a bypass should be done originating from the aortic arch to the arteries distal to the disease to improve blood flow and prevent a stroke if one of the narrowed arteries suddenly occludes.

In the past, the author was reluctant to recommend PTA for treating patients with involvement of the major arteries of the aortic arch. However, more recently, PTA and stenting appear promising. Therefore, it would be advisable to consider PTA particularly for lesions involving

either the right or left common carotid artery. These lesions tend to be long, tapered narrowings that can be treated with PTA and stent placement.

## III. LOWER EXTREMITIES

The abdominal aorta below the diaphragm is the artery most commonly involved in patients with TD. The distribution of the disease is quite peculiar.<sup>9</sup> It usually begins below the renal arteries and then ends just above the aortic bifurcation. There is a unique sparing of the distal aorta. These patients can develop severe claudication, which is poorly tolerated in these young patients. Since the development of disease in the abdominal aorta is slow over a period of time, collaterals frequently develop. Therefore, it is not common to have severe ischemic symptoms such as rest pain, ulcerations, or gangrene.

The patient with abdominal aortic involvement presents interesting challenges. Most abdominal bypass operations for patients with atherosclerosis begin in the abdominal aorta below the renal arteries and go to either both femorals or iliac arteries. However, since the abdominal aorta below the renal arteries is frequently involved with TD, it is inappropriate to originate a bypass from that part of the aorta. The aorta between the diaphragm and the renal arteries is possible as an inflow source but is technically difficult. The author recommends that the proximal anastomosis begin at the thoracic descending aorta and the distal anastomosis to the left iliac artery. Since the distal part of the aorta is spared, a bypass to one iliac artery can perfuse both legs. The procedure is all done retroperitoneally, which is tolerated well with excellent relief of symptoms and good long-term results.

## IV. UPPER EXTREMITIES

The axillary and subclavian arteries are commonly involved with TD, reducing blood flow to both upper extremities. Since muscle mass in the upper extremities is relatively small, symptoms related to exercise of the upper extremities are unusual, as is gangrene and ulcerations in the hands. A significant problem, however, in patients with involvement of the subclavian or axillary arteries is the inability to obtain reliable blood pressure measurements. Hypertension is one of the frequent medical complications of TD, usually due to renal artery or mid-aortic involvement. But with both arms involved with TD, it is impossible to obtain reliable blood pressures, and therefore the diagnosis and treatment of patients with hypertension is a major problem. An attempt to revascularize the upper extremities in order to obtain reliable measurements of blood pressure is a major indication for intervention.

The subclavian steal syndrome can occur in patients with occlusion of the subclavian arteries proximal to the origin of the vertebral artery. Blood is "stolen" from the brain because it flows retrograde down the vertebral artery to supply the arm. Although the incidence of subclavian involvement is high, the "steal" syndrome is unusual because involvement of the arteries is proximal and distal to the origin of the vertebral arteries. But more recently, a few isolated cases of subclavian steal have been reported.<sup>10</sup>

Fortunately, PTA of the subclavian or axillary arteries is becoming more common, particularly with the availability of stents. This procedure is relatively easy to per-

form and seems to yield overall good results.<sup>6</sup> If percutaneous transluminal angioplasty is not possible, then revascularization of the upper extremities should be performed.

## V. RENAL ARTERY DISEASE

Hypertension secondary to renal artery disease is common. The morbidity and mortality from TD frequently is due to unrecognized and untreated hypertension. Congestive heart failure, left ventricular hypertrophy, and myocardial infarction can occur in these relatively young patients. Therefore, any patient with TD and hypertension must have the renal arteries evaluated. Magnetic resonance angiography and Duplex scan of the abdominal aorta and its branches can be an appropriate screening device.

Percutaneous transluminal angioplasty is now the preferred method for treating patients with renal artery stenosis.<sup>11-14</sup> Unlike other arteries involved with TD, the renal artery usually has a short segmental lesion. Therefore, the angiographer would have relatively little difficulty in placing a balloon in that area, dilating it, and then placing a stent to guarantee success of the dilatation. In the past, the results of renal artery dilatation were questionable because of the frequency of restenosis and recurrence. However, the use of the stents may be helpful in reducing the overall incidence of restenosis, and this method can revascularize both renal arteries with improvement in blood pressure.

If percutaneous transluminal angioplasty cannot be performed or is unsuccessful, then surgical bypass is indicated. Usually, bypasses of renal arteries originate from the abdominal aorta below the renal arteries. However, if this is involved with TD, this approach is not possible. The hepatic and/or the splenic arteries have been used as inflow to either renal artery as a method to provide improved blood flow and reduce the incidence of hypertension.

## VI. ANEURYSMS

Aneurysms are relatively uncommon in patients who are born and raised in the United States. This is different than experiences in both Japan and India in which abdominal and thoracic aortic aneurysms are relatively common.<sup>15,16</sup> It is not quite clear why there is such a difference. The incidence of rupture of either the abdominal or thoracic aortic aneurysms is not known. It would appear

to be quite low on the basis of published reports. However, one must understand that these young patients may have 40 to 50 more years of life expectancy, and therefore the risks of aneurysm rupture over that period of time might be quite significant.

Repair of abdominal and thoracic aortic aneurysms is indicated if they reach sizes greater than 5 cm. The new endovascular approach may provide an interesting alternative to patients with abdominal aortic aneurysms from TD. Currently, the author is not aware of any successful placement of endovascular grafts in patients with either abdominal or thoracic aortic aneurysm from TD.

Aneurysms also occur in the innominate and carotid arteries. This presents technical problems, since resection and replacement of these arteries can be difficult with a relatively high incidence of stroke. These cases must be handled individually depending upon the extent of involvement and the symptoms.

## VII. SURGICAL RESULTS

Surgical results for operations on patients with TD are good. There is a very low morbidity and mortality. There is always a concern of the development of anastomotic aneurysms in arteries that have been used for either inflow or outflow bypasses. The author's experience has been that anastomotic aneurysms are relatively uncommon.<sup>8</sup> Constant surveillance of patients who have undergone these procedures is necessary. Anastomotic stenoses, however, do develop. It is not quite clear whether they are due to the recurrence of TD at the level of the anastomosis or other reasons.<sup>8</sup> Nevertheless, they can be treated with either another surgical procedure or possibly even dilatation.

## VIII. CONCLUSION

Takayasu's disease is a difficult problem confronting the vascular surgeon. Most vascular surgeons have very little experience with this entity. It would not be unusual for a surgeon to see no more than one or two patients with this disease throughout his or her entire career. Nevertheless, the vascular surgeon does possess the skills and the experience necessary to provide the appropriate procedure with expectation of good results in those patients who have serious problems due to TD. However, he or she must be aware of the differences between patients with TD and those patients with atherosclerosis.

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