Surgical repair of visceral artery occlusions in Takayasu's disease


We describe the surgical management of six patients with occlusive disease of the visceral arteries caused by Takayasu's arteritis. All patients suffered from renovascular hypertension and, in addition, three of the patients also had symptoms of mesenteric angina. Surgical repair is recommended in the "burn-out" phase of the disease when further progression of the disease is unlikely. Revascularization of the kidneys was by autotransplantation to the iliac vessels. Mesenteric or celiac artery inflow was provided by an extra-anatomic vein bypass graft from the iliac artery. In all cases distal disease-free vessels were available for reconstructive surgery. (J VASC SURG 1986; 3:904-10.)

Inflammatory arterial disease of the Takayasu type is a distinct entity with characteristic pathologic features and age and sex distribution. Although cases have been reported worldwide, this particular type is rare as a cause of vascular disease in western countries but common in the Orient. The disease affects children and young adults and most series report a marked female preponderance.

The pathologic process is a "panarteritis" that initially affects the elastic tissue in the aorta, which is disrupted and surrounded by granulomas with giant cells. The intima responds by proliferation with subsequent thrombosis and the end result is stenosis, occlusion, or aneurysm formation. Commonly one or more of the major branches from the aortic arch and abdominal aorta are occluded at their origin.

Aortography reveals diffuse irregular involvement of the aortic arch and the descending and abdominal aorta. A smaller group of patients also has involvement of the pulmonary artery.

The disease may start as a nonspecific "rheumatic-like" illness with rashes, fever, arthralgia, anemia, and an elevated erythrocyte sedimentation rate (ESR) but this phase is not recognized in most patients. Some years later, patients show the sequelae of their vascular occlusive disease, which can include arm claudication, renal hypertension, and abdominal angina. Takayasu's disease can be distinguished from other inflammatory diseases that involve the aorta and its major branches, such as tuberculosis, syphilis, giant cell arteritis, polymyalgia rheumatica, and the inflammatory variant of atheroma.

MATERIAL AND METHODS

This report describes the surgical treatment and outcome (Table I) in six patients who presented with severe hypertension and, when investigated, were shown to have Takayasu's arteritis of the aorta with occlusion of one or more of the visceral branches of the abdominal aorta.

CASE REPORTS

Case 1. The patient was a 12-year-old Chinese girl who initially came to the hospital with fever, oliguria, hematuria, and peripheral edema. A renal biopsy showed a proliferative glomerulonephritis. Evidence of nephritis slowly resolved over a 6-month period while the patient was treated with steroids. She was readmitted 5 years later, aged 17 years, complaining of headaches and vomiting. She was found to be severely hypertensive; blood pressure was 220/140 mm Hg in the right arm and 160/130 mm Hg in the left arm. She had a left renal bruit, grade II retinopathy, and moderate cardiomegaly.

Studies undertaken in hospital included microscopic examination of the urine, which was normal, urinary protein (0.1 gm/24 hr), and creatinine clearance (38 ml/min). An intravenous pyelogram (IVP) showed prompt bilateral function of the kidneys and the aortogram revealed diffuse aortic involvement with occlusion of the left subclavian artery, occlusion of the right renal artery, and severe ste-
nosis of the left renal artery (Fig. 1). Renal vein renin estimations were elevated above the normal range on both sides. Surgery was undertaken to correct occlusive disease of the renal arteries. A right aorto–renal vein bypass graft and left autotransplantation was performed.

A full-thickness biopsy of the aortic wall was taken at the site of the anastomosis and demonstrated the characteristic histologic features of Takayasu's disease (Fig. 2, A and B) with destruction of the elastic fibers in the media and chronic inflammatory cell infiltrate in all layers with granulomas and characteristic multinucleate giant cells and an absence of caseation.

Postoperatively she had an initial prompt diuresis and resolution of hypertension. Twelve hours after operation severe bleeding developed. At laparotomy there was diffuse bleeding, but no specific bleeding points were identified. Platelet counts showed severe thrombocytopenia that persisted for 2 weeks. Acute kidney failure developed that required dialysis for 4 weeks. Hematologic studies and renal biopsy supported the diagnosis of thrombotic thrombocytopenia. Slow improvement in renal function allowed the patient to survive without dialysis, but she suffered markedly impaired renal function. Aortography confirmed patent revascularization of both kidneys. The patient died of kidney failure 6 months after operation.

Case 2. A 15-year-old Malay boy came to the hospital complaining of headaches and vomiting of 1 month's duration. He was hypertensive, with a blood pressure of 160/120 mm Hg in both arms. The fundi were normal and a loud bruit was heard in the epigastrium. Physical examination gave otherwise normal findings. Laboratory tests indicated a normal serum creatinine level and a mildly elevated ESR (17 mm/hr). An IVP indicated no abnormalities and some aortic calcification was noted. Aortography demonstrated bilateral renal artery stenosis, marked stenosis of the celiac and superior mesenteric arteries, and grossly irregular thoracic and abdominal aorta. At operation he had bilateral renal autotransplantation. An IVP showed normal function of both kidneys 2 weeks after operation. He was normotensive when discharged from the hospital. A repeat aortogram 1 month later showed excellent revascularization to both kidneys, and blood pressure was 110/70 mm Hg.

He was readmitted to the hospital 6 months later with severe abdominal pain and, at laparotomy, widespread bowel infarction with perforation and soiling of the peritoneum was found. He died 12 hours after operation. No autopsy was performed.

Case 3. The patient was a 9-year-old Malay boy. He had been in the hospital many times during the previous 12 months, suffering from cardiac failure and poorly controlled hypertension, despite appropriate medical management with digoxin, diuretics, and beta-blockers. He was thin and malnourished and gave a history suggestive of mesenteric angina. Clinical examination showed a grossly enlarged heart and a loud epigastric bruit. Fundi were normal. Hemoglobin, white blood cell count, ESR, and serum creatinine values were in the normal range. An IVP demonstrated normal function on the right, but delayed flow into the celiac circulation established by a saphenous vein graft from the left iliac artery to the splenic artery (Fig. 3). Blood supply to the abdominal viscera came from a dilated inferior mesenteric artery, filling via a grossly enlarged marginal artery of Drummond to the mesenteric and celiac circulation.

Surgical repair undertaken in this young boy involved bilateral autotransplantation of the kidneys and arterial inflow into the celiac circulation established by a superficial femoral bypass graft from the left iliac artery to the splenic artery (Fig. 4). The spleen was not removed. After operation, renal function and blood pressure were normal. His exercise tolerance improved and he soon put on weight and could enjoy vigorous sport at school. A postoperative arteriogram at 6 months (Fig. 5) when he was hypertensive (140/90 mm Hg) demonstrated dilatation of the iliosplenic bypass and excellent revascularization of the celiac and mesenteric arteries.

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Table I. Surgical treatment and outcome for six patients with Takayasu's disease

<table>
<thead>
<tr>
<th>Case</th>
<th>Year (yr)</th>
<th>Sex</th>
<th>Visceral artery disease</th>
<th>Operation</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1974</td>
<td>F</td>
<td>L RA stenosis; R RA occlusion</td>
<td>L autotransplant R aorto-renal vein bypass graft</td>
<td>Died 6 mo postop of kidney failure</td>
</tr>
<tr>
<td>2</td>
<td>1978</td>
<td>M</td>
<td>Bilateral RA stenosis; celiac stenosis; superior mesenteric stenosis</td>
<td>Bilateral autotransplant</td>
<td>Died at 6 mo of bowel infarction</td>
</tr>
<tr>
<td>3</td>
<td>1979</td>
<td>M</td>
<td>Bilateral RA occlusion; celiac and superior mesenteric occlusion</td>
<td>Bilateral autotransplant; iliosplenic bypass graft</td>
<td>Mild hypertension; no drugs</td>
</tr>
<tr>
<td>4</td>
<td>1980</td>
<td>M</td>
<td>R RA stenosis</td>
<td>R autotransplant</td>
<td>Normotensive; no drugs</td>
</tr>
<tr>
<td>5</td>
<td>1983</td>
<td>M</td>
<td>R RA occlusion; L RA stenosis; celiac and superior mesenteric occlusion</td>
<td>Bilateral autotransplant; ilio-superior mesenteric bypass graft</td>
<td>Normotensive; no drugs</td>
</tr>
<tr>
<td>6</td>
<td>1984</td>
<td>M</td>
<td>Bilateral RA occlusion</td>
<td>Bilateral autotransplant</td>
<td>Normotensive; no drugs</td>
</tr>
</tbody>
</table>

L = left; R = right; RA = renal artery.
Fig. 1. Arteriogram of case 1 (17-year-old female patient) shows diffuse irregularity of abdominal aorta with occlusion of the right renal artery and severe stenosis of the left renal artery. Vessels distal to aortic bifurcation were normal.

case 5. A 25-year-old Malay man came for a medical examination to enter the Army. He was hypertensive and after close questioning, he remembered abdominal pains occurring after large meals, suggestive of mesenteric angina. He was a muscular, well-nourished man who had been working on a rubber plantation. He had a loud abdominal bruit and a palpable epigastric thrill. Laboratory studies showed normal values of hemoglobin, white blood cell count, and ESR and creatinine clearance of 50 ml/min. Aortography (Fig. 6) demonstrated diffuse involvement of the aorta, occlusion of the celiac, superior mesenteric, and right renal artery, and stenosis of the left renal artery. Mesenteric vessels filled from the grossly dilated inferior mesenteric artery and collateral vessel of Drummond. Surgical reconstruction in this case involved bilateral autotransplantation and iliac artery to superior mesenteric artery bypass with the use of saphenous vein (Fig. 7). After operation he has been normotensive and is no longer receiving drug therapy. Postoperative angiography demonstrated both renal autotransplants and a patent iliofemoral bypass. This patient has remained normotensive in the 2 years since operation.

Case 6. A 19-year-old male patient initially came to the hospital with a right-sided stroke that developed suddenly at work. He was found to be severely hypertensive (160/120 mm Hg). The ESR was 19 mm/hr. Renal function was normal and intravenous pyelography was normal with vascular notching of both ureters. Aortography demonstrated an irregular abdominal aorta with bilateral renal artery stenosis of the proximal renal artery and poststenotic dilatation. He was initially treated medically but with poor control until captopril was introduced. Six months after the onset of his stroke when disability was minimal, he underwent bilateral renal autotransplantation. After operation he had normal renal function and blood pressure without drug therapy. Intravenous pyelography confirmed the function of both kidneys after the operation. Twelve months after operation the patient remains normotensive without hypertensive drug therapy.

DISCUSSION

In this series of patients with Takayasu's disease undergoing surgical reconstruction, only one patient gave a definite history of an initial acute or subacute "rheumatic-like" illness. As all of these patients came from a relatively poor socioeconomic background in a developing country in which minor illness is frequent and often not reported to medical practitioners, it is likely the medical history was incomplete.

The origin of this disease is unknown. The pathogenetic mechanism appears to be destruction of the elastic tissue of the aortic wall with development of granulomas with multinucleate giant cells without caseation, which represents a histologic picture also seen in polymyalgia rheumatica and giant cell arte-
Fig. 2. Case 1. A, Photomicrograph of aortic wall demonstrates irregular destruction of elastic fibers in the media (elastic van Gieson stain; original magnification, ×30). B, This photomicrograph demonstrates area of chronic inflammatory cells arranged as a granuloma with multinucleate giant cells without caseation in wall of aorta (hematoxylin-eosin stain; original magnification, ×120).

The disease process is thought to have an initial acute, then subacute phase with the hallmarks of a systemic illness, diffuse pains, anemia, and an elevated ESR. During this phase of the disease, steroids may play a role in reducing the activity. Surgery should not be performed until the disease is in a "burn-out" phase with a normal ESR. It is at this phase of the disease that patients show sequelae of their vascular occlusive disease, such as hypertension or claudication.

Should the sequelae be of such severity to warrant investigation, aortography is necessary to document fully the extent of the disease and the possibility of...
reconstruction. In this series, all cases showed diffuse involvement of the thoracic and abdominal aorta but were sparing of the iliac and more distal arteries. In each case in which the kidneys were autotransplanted to the iliac arteries, histologic examination biopsies taken at the anastomotic region of the renal artery and iliac artery yielded normal findings. The disease process was restricted to the wall of the aorta and the branches as they passed through the aortic wall. In the follow-up period to date, we have no evidence that the disease process has reactivated to involve more distal arteries.

Investigation of hypertension, particularly in young female patients, may suggest fibromuscular disease and aortography showing occlusion of the renal artery suggests that possibility. However, the addition of diffuse irregular involvement of the thoracic and abdominal aorta allows a confident diagnosis of Takayasu's arteritis. The other frequent vascular disease in the Orient is thromboangiitis obliterans, which is seen in young and middle-aged adults who are almost invariably smokers. This disease process results in digital ischemia and gangrene and occasionally in a major amputation. Arteriography again is distinctive with normal major vessels and occlusion of vessels distal to the popliteal and brachial arteries. Characteristic "corkscrew" collateral branches are not unique to this condition but testify to the chronicity in relatively young patients. This more peripheral pattern of disease should not be confused with Takayasu's arteritis. In each of the three conditions, the histologic pattern is sufficiently characteristic and different to allow a firm pathologic diagnosis from a biopsy of an involved artery.

The surgical approach in each case has been to attempt a reconstruction away from the diseased aorta. Autotransplantation of the kidney is a well-established procedure. If both renal arteries are involved, it has been our practice to do a bilateral autotransplantation at the one operation. If the renal artery is occluded, there is usually no information from the aortogram whether single or multiple renal arteries are present, so the nephrectomy should be done cautiously and one must be prepared to reconstruct multiple renal arteries by ex vivo bench surgery if necessary. We have done vascular reconstruction on the left side to the internal iliac artery and external iliac vein and on the right side to the common iliac artery and common iliac vein. This procedure allows one of the internal iliac arteries to remain patent. The ureters are not divided and are only mobilized enough to lie without tension or kinking. Operation
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Fig. 5. Arteriogram in case 3, 6 months after operation, shows bilateral renal autotransplants and considerable dilatation of saphenous vein graft from iliac to splenic artery. This study also demonstrates a stenosis of the artery to left renal autotransplant.

Fig. 6. Aortogram of patient 5 (25-year old man) demonstrates complete occlusion of celiac, superior mesenteric, and right renal artery and stenosis of the left renal artery. Also shown is grossly dilated collateral branch from the inferior mesenteric artery to the superior mesenteric artery.

Fig. 7. This diagram illustrates surgical reconstruction in case 5. Renal artery repair was by bilateral autotransplantation. Mesenteric inflow was achieved by left common iliac-to-superior mesenteric artery vein bypass graft.

has been performed through a long midline incision and the patients have not been heparinized.

Patient 2 who died of bowel infarction 12 months after operation highlighted the hazard of celiac and mesenteric ischemia, particularly when hypertension is reduced. Cases 3 and 5 with symptoms of mesenteric angina and celiac and superior mesenteric occlusions demanded a further surgical reconstruction. In the small child the left iliac to splenic artery reconstruction appeared, at the time, a relatively simple solution and has worked well. The left iliac artery-to-superior mesenteric artery bypass in case 5 is probably a better surgical approach because it is not dependent on the celiac to superior mesenteric anastomotic network in which the superior mesenteric supply area is the more frequent site of bowel ischemia and infarction.

Takayasu's disease is relatively rare, but it affects young patients who may survive for many years. We conclude that reconstructive vascular surgery in the "burn-out" phase of the disease is appropriate, as patients can live for many years without further progress of the arterial occlusive process.

REFERENCES
3. Daneraj TJ, Wong HO, Thomas MA. Primary arteritis of