Surgical Treatment for Cardiac Lesions Combined with Takayasu's Aortitis

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ABSTRACT. Surgical treatment for cardiac lesions combined with Takayasu's aortitis was carried out in six cases; four with aortic regurgitation, one with bilateral coronary ostial stenosis, and one with both aortic regurgitation and right coronary ostial stenosis. Active inflammation was seen in three cases and was treated with prednisolone pre- and postoperatively. Three cases had stenoses of the branches of the aortic arch and one of these with aortic regurgitation and hypertension died of brain damage due to intraoperative low cerebral perfusion. Aortic valve detachment occurred in one case with active inflammation, and reoperation was done 9 and 15 months after the initial operation. Transaortic coronary ostial endarterectomy was performed on three coronary arteries in two cases and sufficient patency was confirmed angiographically after surgery.

Key words: Takayasu's aortitis — aortic regurgitation — coronary ostial stenosis

It is well known in patients with Takayasu's aortitis that the clinical features and prognosis vary with the location, spread and degree of the lesion in the affected artery, but the most common cause of death in Takayasu's aortitis is cardiac disease. In Japan, 26% of 744 cases that were treated surgically were operated on for combined cardiac lesions; i.e., aortic valve replacement in 155 cases and coronary reconstructive surgery in 48 cases.[1] We treated six cases surgically for aortic regurgitation and/or coronary artery ostial stenosis combined with Takayasu's aortitis. These cases and some problems we encountered in our operative procedures are discussed in the present paper.

CASES

Between 1975 and 1993, we experienced 41 cases of Takayasu's aortitis at Kawasaki Medical School Hospital. Of these, six (14.6%) underwent surgical treatment for combined cardiac lesions; Four for aortic regurgitation, one for bilateral coronary ostial stenosis and one for both aortic regurgitation and right coronary ostial stenosis. All the patients were female, and the cases of aortic regurgitation ranged in age from 38 to 51 years old. The patient with bilateral coronary stenosis was 17 years old and the one with both aortic regurgitation and right coronaty stenosis was 38 years old. The duration of illness ranged

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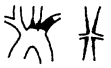
from 2 to 23 years in the cases of aortic regurgitation. In the two cases with coronary stenosis, however, the duration was only two and three months, respectively. Active inflammation was seen in three cases and was suppressed sufficiently with prednisolone pre- and postoperatively. Patient profiles of these six cases are presented in Table.

TABLE Surgical cases of cardiac lesions combined with Takayasu's aortitis
AR : aortic regurgitation AVR : aortic valve replacement SA : subclavian artery
LCA : left common carotid artery PVL : perivalvar leakage

Case	Age Sex	Diagnosis	NYHA	Duration of Illness	Inflammation	Angiogram	Operation	Results
1	38 f	AR 3/4	3	14 ys	—		AVR	SA-LCA bypass alive
2	49 f	AR 3/4 hypertension	4	23 ys	—		AVR	brain damage died
3	49 f	AR 3/4	3	2 ys	—	—	AVR	alive
4	51 f	AR 4/4	3	7 ys	active		AVR	alive
5	17 f	angina	4	2 ms	active		endarterectomy	alive
6	38 f	angina AR 3/4	4	3 ms	active		AVR endarterectomy	PVL reope re-reope

1 Aortic regurgitation

Five of our cases, including the case with right coronary ostial stenosis, had aortic regurgitation with the severity of the regurgitation being grade 3 in four cases and grade 4 in the remaining case. Mild or moderate dilatation of the ascending aorta was seen in aortograms in all cases and stenosis of the branches of aortic arch was seen in three cases. One of these had an atypical coarctation of the abdominal aorta with hypertension. At the time of operation, the ascending aorta was tough and thickened with edematous intimal thickening and the aortic valve was also edematous with fibrous thickening. The annular dilatation was not so marked as to become the main cause of aortic regurgitation. The aortic valve was replaced using a 23 mm St Jude Medical artificial heart valve by buttressed suture with a spaghetti pledget in all cases. Histological examination of the excised aortic cusps showed fibrous myxomatous degeneration and no inflammatory findings even in two cases with active inflammation in the aortic wall. Perivalvar leakage occurred six months after aortic valve replacement in one case whose inflammatory reaction was well suppressed with prednisolone, and two perivalvar defects found in the sinus of Valsalva (Fig. 1) were closed tightly by buttressed suture with a large Teflon pledget nine months after the initial operation. Three months after the second operation, this patient had a recurrence of perivalvar leakage, and reoperation was performed six months after the second operation because of rapid progression of aortic regurgitation. There were multiple perivalvar defects and an unstable prosthetic valve was observed (Fig. 2). The prosthetic valve was removed and the aortic root was completely replaced using a valved conduit employing Cabrol's methods[2] successfully (Fig. 3).

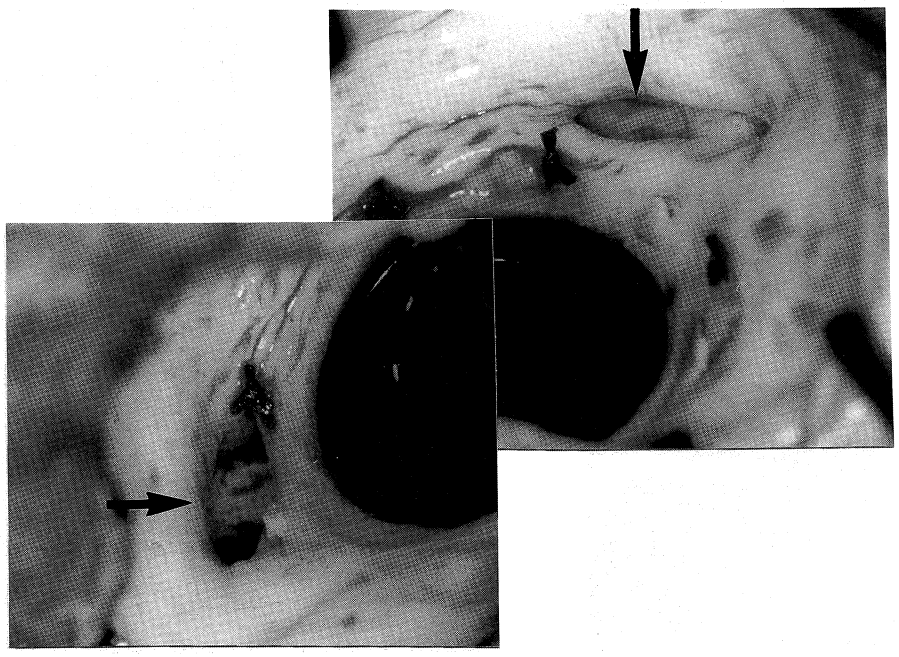


Fig. 1. Two Perivalvar detachments (arrows) in the sinus of Valsalva in a 38-year-old woman with perivalvar leakage nine months after aortic valve replacement

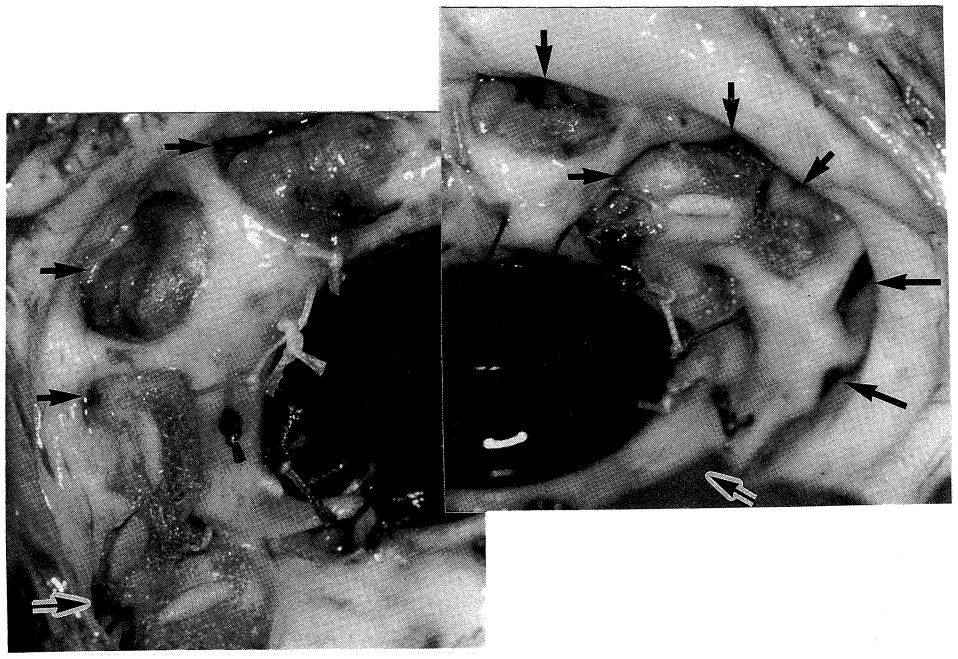


Fig. 2. Recurrence of perivalvar leakage three months after the second operation Multiple small perivalvar defects (arrows) and an unstable prosthetic valve were observed.

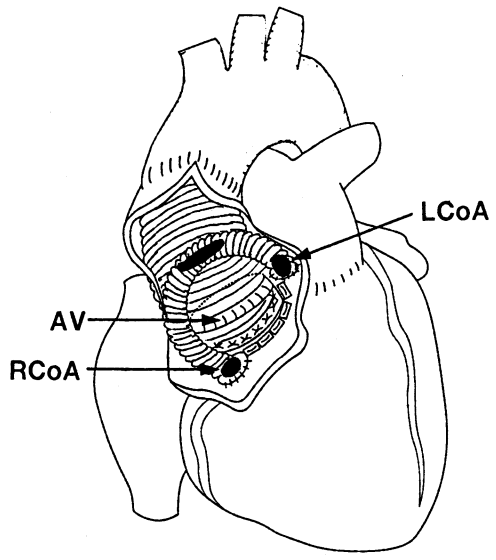


Fig. 3. Replacement of the aortic root with translocated valvar conduit and reimplantation of coronary arteries employing Cabrol's method
AV : artificial heart valve RCoA : right coronary artery LCoA : left coronary artery

Brain damage considered to be due to low cerebral perfusion during cardiopulmonary bypass occurred in one case with hypertension and stenosis of both the left carotid and left subclavian arteries. This patient died of subarachnoideal hemorrhage six months after the operation.

2 Coronary ostial stenosis

Transaortic coronary ostial endarterectomy (Fig. 4) was performed in three coronary arteries of two cases, a 17-year-old girl with bilateral coronary ostial stenosis and a 38-year-old female with right coronary ostial stenosis and aortic regurgitation. Active inflammation was seen in both cases, and it was treated

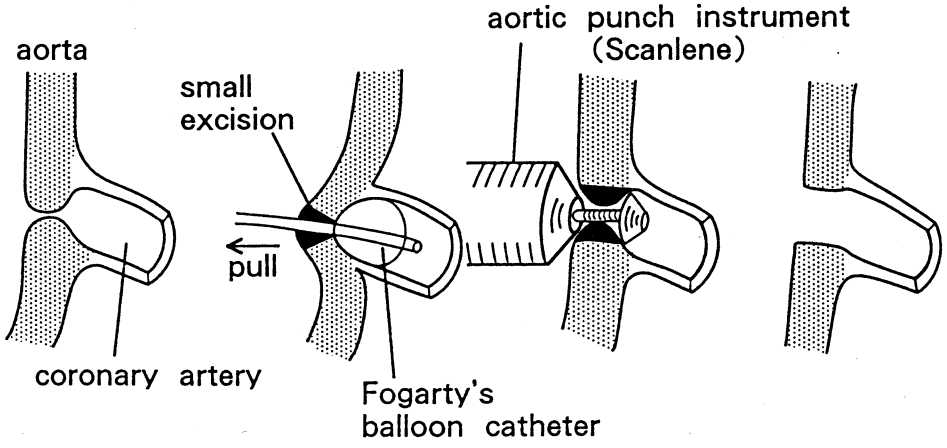


Fig. 4. Operative procedures of transaortic coronary ostial endarterectomy

with prednisolone before and after the operation. Postoperative coronary angiograms revealed sufficient enlargement of the coronary orifices (Figs. 5-7). In the first case, follow-up coronary angiograms nine years after the operation showed sufficient patency and no progression of stenosis in the left and right coronary orifices. In the second case, a good enlargement of the right coronary orifice was confirmed during the first and second reoperation for perivalvar leakage.

DISCUSSION

The reported incidence of aortic regurgitation combined with Takayasu's aortitis in Japan is unexpectedly high; 33.3% of the 1307 cases registered in Japan in 1992.[3] The etiology of the aortic regurgitation in Takayasu's aortitis

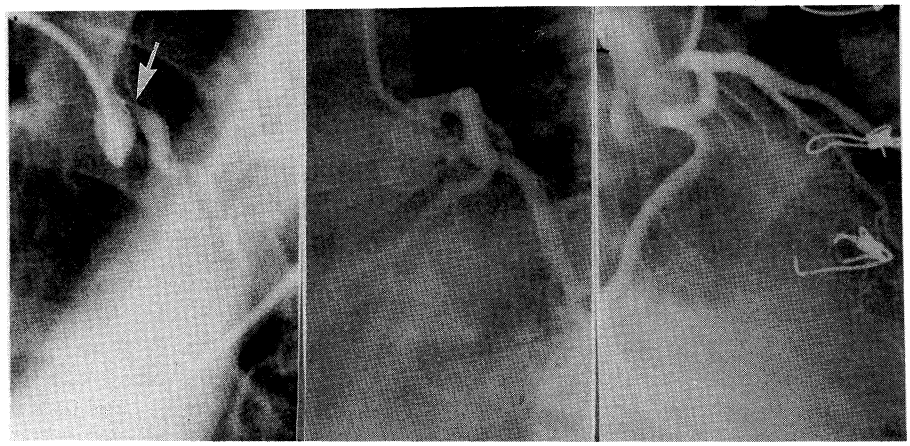


Fig. 5. Left coronary arteriograms before the operation (A) and two months (B) and 9 years (C) after transaortic coronary ostial endarterectomy in a 17-year-old girl

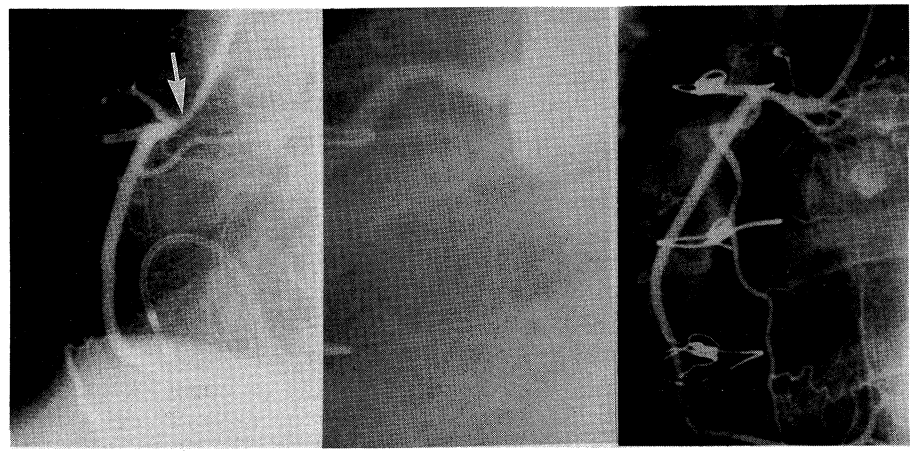


Fig. 6. Right coronary arteriograms before the operation (A), and two months (B) and nine years (C) after transaortic coronary ostial endarterectomy in the same patient

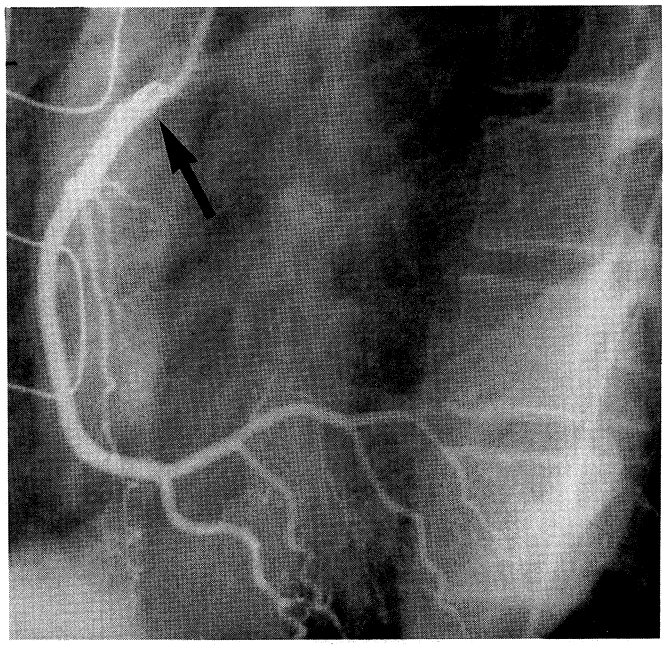


Fig. 7. Right coronary arteriogram two months after transaortic coronary ostial endarterectomy in a 38-year-old female with aortic regurgitation

is 1) organic deformity due to inflammation of the aortic cusp, 2) annulo-aortic ectasia. However, in five of our cases, the annular dilatation was not so marked as to become the main cause of aortic regurgitation. In addition, histological examination of the excised aortic cusps showed fibrous myxomatous degeneration and no inflammatory findings even in cases with active inflammation in the aortic wall. These results suggest that the myxomatous degenerative changes in the aortic cusp and the annular ring of the sinus of Valsalva may be attributable to the development of aortic regurgitation in Takayasu's aortitis.

Late aortic valve detachment is one of the noteworthy complications after aortic valve replacement in Takayasu's aortitis. Although it has been noted that active inflammation is one of the main factors of aortic valve detachment,[4,5] as seen in our patient, it may occur even in a case in which inflammation has been sufficiently suppressed by steroid therapy. Therefore, it is considered that myxomatous degeneration of the annular ring and prednisolone administered for active inflammation may affect healing of the valve replacement. To prevent aortic valve detachment, it may be necessary to fix the artificial heart valve from the outside of the aortic wall with a large pledget. In the surgical management of aortic valve detachment, aortic root replacement using a valved conduit with a translocation of the prosthetic valve in the vascular graft and coronary reimplantation would be necessary to abate the diastolic stroke to the suture line.[6]

Systemic hypertension and obstructive lesions in a branch of the aortic

arch are frequently seen in Takayasu's aortitis, and it is important to maintain high flow and high pressure perfusion while performing hypothermic cardiopulmonary bypass to prevent brain damage due to low cerebral perfusion in these cases.

Approximately 10% of cases with Takayasu's aortitis involve the ostia of the coronary artery,[7] which causes fatal cardiac complications. As a result, many of these patients become candidates for surgery. As for the surgical procedures, Ohara *et al*[8] reported that coronary bypass surgery was indicated for patients with total occlusion of the coronary artery ostia or with peripheral coronary lesions and that a transaortic coronary ostial endarterectomy was strongly indicated in cases with a marked thickening of the aortic wall and with stenosis localized at the ostia. In 33 surgical cases compiled by Yamazaki *et al*,[9] 38 aortocoronary saphenous vein bypass graftings were performed and a transaortic coronary ostial endarterectomy was performed in only 7 cases. In the literature, bypass grafting appears to be most commonly used for coronary ostial stenosis. Compared with the patency rate of vein grafts in arteriosclerotic coronary heart disease, the rate reported for aortitis syndrome is low.[8] In addition, occlusion is frequently seen in the proximal anastomosis of the graft.[8] This may be caused by technical difficulty in the anastomosis of the vein graft to the thickened and sclerotic aortic wall. Furthermore, active inflammation of the aortic wall also may affect the graft occlusion. Recently, use of coronary ostial patch angioplasty was reported in the literature,[10] but the long-term results of this procedure remain unclear. Restenosis due to intimal reproliferation may be the most likely complication to occur after coronary ostial endarterectomy for Takayasu's aortitis. However, with the exception of the case we have reported[11] there have been no reports of long-term follow-up angiography following this procedure. While it is possible to suppress the inflammatory reaction with steroid therapy, whether or not progression of the lesions is stopped remains unknown, and it is unclear what effect postoperative steroid therapy has in suppressing the development of intimal reproliferation. Transaortic coronary ostial endarterectomy is considered to be an anatomically reasonable operation for coronary ostial stenosis combined with Takayasu's aortitis because long-term patency may be anticipated.

In conclusion, some problems were experienced in the surgical management of cardiac lesions in Takayasu's aortitis; namely, valve detachment after aortic valve replacement and brain damage due to low cerebral perfusion during cardiopulmonary bypass. The operative procedures for coronary ostial stenosis have been presented.

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