Congenital Coronary Artery Fistula
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Surgical Results and Late Changes in Coronary Artery Aneurysm

Haruo Miyamura, M.D., Shoji Eguchi, M.D., Hiroshi Watanabe, M.D.
Masaaki Sugawara, M.D., Yoshiki Takahashi, M.D.
Mayumi Shinonaga, M.D. and Shoh Tatebe, M.D.

Four pediatric cases of congenital coronary artery fistula were surgically treated and followed for 8 years. In the 3 cases of right coronary artery to right ventricle fistula, regression of coronary artery dilatation was observed postoperatively. In the 1 case of circumflex artery to right atrium fistula, aneurysmal dilatation of the abnormal vessel persisted for 8 years. A reduction in vessel size is expected if the fistula-related coronary artery has a normal course and normal branchings. When the aneurysmal vessel takes an abnormal course without branches, it should be removed surgically along with fistula closure. (Jpn Circ J 1995; 59: 786–789)

Congenital coronary artery fistula (CCAF) is a communication between a coronary artery and one of the cardiac chambers, the superior vena cava, or a pulmonary artery. It may cause congestive heart failure, infective endocarditis, and/or coronary artery aneurysm! A small fistula is not usually treated surgically. However, if a moderate to large amount of shunting is confirmed, closure of the fistula is indicated. The coronary artery proximal to the fistula is usually massively dilated, but surgical treatment of this vessel is controversial. Some authors have recommended plication of the dilated portion of the artery? However, spontaneous regression of the dilatation has been reported after closure of the fistula? In 1986, 4 pediatric cases of CCAF underwent surgery at our institute. We report here the surgical procedures and their late results, with a particular emphasis on morphological changes in the dilated coronary artery.

Key words:
Congenital coronary artery fistula (CCAF)
Coronary artery aneurysm
Fistula closure
Dilated coronary artery

PATIENTS AND METHODS

Four children with CCAF were referred to our department for surgical repair in 1986. Preoperatively, all were asymptomatic, however, upon auscultation a loud continuous murmur was heard at the precordium. Their ages, sex, and hemodynamic data are listed in Table I. In 3 cases (Patients 1, 2, 3), CCAF existed between the right coronary artery and the right ventricle, and in 1 case (Patient 4) a fistulous vessel ran between the circumflex artery and the right atrium. All of the coronary arteries involved showed massive dilatation. The operation was performed through a median sternotomy in all cases. While the patient was under moderate hypothermic extracorporeal circulation, the aortic root was cross-clamped, and cardiac standstill was obtained with cold cardioplegic solution. For the 3 cases of right coronary artery to right ventricle fistula (Patients 1, 2, 3), the dilated coronary artery just above the fistula was cut open, and the communication was closed by pledged sutures. The incised coronary artery was then closed.

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Department of Thoracic and Cardiovascular Surgery, Niigata University School of Medicine, Niigata, Japan
Mailing address: Haruo Miyamura, M.D., Department of Thoracic and Cardiovascular Surgery, Niigata University School of Medicine, Asahi-machi 1-757, Niigata City, 951, Japan

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TABLE I SUMMARY OF PATIENTS' DATA

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age at Operation (yr)</th>
<th>Sex</th>
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<td>9</td>
<td>F</td>
<td>CX</td>
<td>RA</td>
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RCA: Right coronary artery, RV: Right ventricle, CX: Circumflex artery, RA: Right atrium, PAP: Pulmonary artery pressure, Qp/Qs: Pulmonary to systemic blood flow ratio, F: Female, M: Male.

Fig.1. Morphological changes in the dilated coronary artery before and 1 year after surgery.

Fig.2. Right coronary arteriography in Patient 3 from the right anterior oblique view. Dilatation of the coronary artery with fistula formation before surgery (A), and the regression of vessel dilatation after surgery (B) are shown.
with sutures. Plication of the coronary artery was not performed, and the diffusely dilated artery was left untouched. For the case of circumflex artery to right atrium fistula (Patient 4), the communication was closed through a right atriotomy and the fistulous vessel was not resected. All 4 cases had an uneventful postoperative course, and were discharged from the hospital. Postoperative angiography was performed one year after the surgery to evaluate morphological changes in the coronary artery.

RESULTS
The patients have been followed at our outpatient clinic for 8 years, and have all been well without any complaints or symptoms. The pre- and post-operative angiography findings are shown in Fig. 1. Regression of coronary artery dilatation was observed in Patients 1, 2, and 3. Fig. 2 shows the angiographic change in the right coronary artery in Patient 3. In Patient 4, the aneurysmally dilated fistulous vessel between the circumflex artery and the right atrium persisted for 1 year postoperatively. This aneurysmal vessel ran posterior to the right and left atrial wall without detaching any branches, and this angiographic finding has not changed for 7 years after the surgery (Fig. 3).

DISCUSSION
The first successful surgical intervention in CCAF was reported by Björck and Crafoord in 1947. However, the surgical indications for this rather rare anomaly have not yet been clearly defined. Jaffe et al. reported a case of spontaneous occlusion of the right coronary artery proximal to the fistula, and questioned the efficacy of the surgery by noting that little anatomic or functional change occurred over a prolonged follow-up period if the shunt was small-to-moderate. On the other hand, Libeithson et al. stated that patients with CCAF who were more than 20 years old had more preoperative symptoms and more postoperative complications. Therefore, they concluded that CCAF should be closed electively during early childhood, even if it was asymptomatic. Bogers et al. also recommended the elective surgical closure of CCAF in asymptomatic patients to prevent complications of the fistula and postoperative problems after surgery. The patients in our series were all asymptomatic preoperatively, but cineangiography showed large fistula communications in which spontaneous closure was very unlikely to occur. Therefore, we performed the surgical closure of CCAF during childhood. Many surgical techniques for closing fistula have been reported, and some maneuvers can be performed without cardiopulmo-
nary bypass! While we believe that each technique offers satisfactory results if properly used, we used cardiopulmonary bypass and cardiac arrest in all cases to achieve accurate closure of the fistula under direct visual observation. Hallman et al reported a technique called “lateral arteriorrhaphy”, which enabled simultaneous fistula closure and plication of the dilated coronary artery by placing multiple mattress sutures tangential to the posterior surface of the coronary artery? We did not use this method, which is technically difficult and carries a risk of coronary artery occlusion. Oldham et al stated that if the fistula communication was properly obliterated at its site of entry into the cardiac chamber, the aneurysmal coronary artery could be expected to return to normal size3 In our series, regression of the massively dilated coronary artery was dramatic in the 3 cases of right coronary artery to right ventricle fistula. In these 3 cases, the course and branching distributions of the fistula-related coronary artery were normal, and we assume that the reduction in flow was responsible for the dilated vessel returning to normal size. Jaffe et al reported that the degree of dilatation of the involved coronary artery was essentially unchanged following surgery. However, the 4 patients they reported (9, 11, 16 and 40 years old) were much older than the patients in our series. Thus, the age distribution of the patients might have affected their result. The 1 case in which we did not observe regression of aneurysmal dilatation of the vessel was the patient with circumflex artery to right atrium fistula. The abnormal fistulous vessel arose from the circumflex artery and entered the right atrium without any branches. Surgical closure of the fistula through the inside of the atrial wall was not associated with any changes in the size of the aneurysmal vessel postoperatively. This abnormal vessel should have been removed or ligated at its origin to prevent the risk of rupture, which is, fortunately, reported to be extremely rare5.

In conclusion, we observed the reduction of coronary artery dilatation after fistula closure in 3 cases in which the fistula-related artery showed a normal course and normal branchings. In the 1 case in which the artery showed an abnormal course without branch- es, aneurysmal dilatation persisted long after surgery, and we assume that this vessel should have been removed or ligated at its origin at the time of fistula closure.

REFERENCES