Background: Few studies after surgical repair of the anomalous origin of the left coronary artery have reported the importance of the mitral annuloplasty or the long-term results.

Methods: Between January 1982 and March 2000, 29 patients with anomalous origin underwent surgical intervention at our institution (direct aortic reimplantation in 19 and Takeuchi procedure in 10). Age at the time of operation ranged from 2 months to 24 years (median, 29.3 months), and 9 patients were infants. Twenty-four patients had varying degrees of mitral incompetence. Simultaneous mitral annuloplasty at the anterolateral commissure was performed in all 24 patients with incompetence.

Results: There were 2 hospital deaths among the infants, and no late deaths. Mean follow-up was 100 ± 57 months, and the actuarial survival was 93.1% at 10 years (70% confidence limits, 87-99). Cardiothoracic ratio at discharge was not decreasing significantly \( (P = .35) \); however, this value 5 years after the operation showed the significant decrease \( (P = .003) \) versus preoperative value. Preoperative mitral incompetence decreased in all but one of the operative survivors with mitral annuloplasty at the last follow-up. The left ventricular fractional shortening z-score was not normalized at discharge but was normalized in the late period.

Conclusion: These data demonstrate that impaired left ventricular function normalized in the long term (even if it was below normal immediately after operation) after 2-coronary repair. We recommend that the simultaneous mitral annuloplasty should be performed at the time of operation for patients who have mitral incompetence with anomalous origin of the left coronary artery.(J Thorac Cardiovasc Surg 2001;121:792-7)
Patients. Between January 1982 and March 2000, 29 patients with ALCAPA underwent surgical intervention at our institution. There were 8 male and 21 female patients. The median age at the time of operation was 29.3 months (range, 2 months-24 years), and 9 patients were less than 12 months old. Congestive heart failure, as defined by the need for diuretics, was present in 15 (52%) patients. Cardiomegaly, defined as a cardiothoracic ratio (CTR) of greater than 0.55, was observed in 22 (76%) patients. The median CTR was 0.61 (range, 0.57-0.74). The electrocardiograms of all 9 infants and 15 of 20 older children showed Q waves in lead I, aVL, or both. The mean preoperative FSz for infants less than 1 year old was –8.1 ± 4.1, and that for older children was –0.6 ± 3.1. Twenty-four (83%) patients had varying degrees of mitral incompetence: mild in 6 (21%); moderate in 11 (38%); and severe in 7 (24%).

Surgical procedures. Operations for all patients were performed through a median sternotomy, with bicaudal cannulation and hypothermic cardiopulmonary bypass. Cardiac arrest was induced by infusion of a crystalloid cardioplegic solution through the aortic root, with crossclamping of both great vessels. Nineteen (66%) patients underwent direct aortic reimplantation, and 10 (34%) patients underwent a Takeuchi procedure. The main pulmonary artery (PA) was transversely opened just proximal to the bifurcation, and the location of anomalous orifice was identified.

Direct reimplantation. The main PA was then divided. The left coronary artery was removed in a button-shaped cuff and was subsequently mobilized. The aorta was opened transversely, and an opening was created into the left coronary sinus. The left coronary button was anastomosed to the aorta from within the aortic lumen by means of 7-0 absorbable continuous sutures. The coronary defect in the PA sinus was closed with an autologous pericardial patch, and PA continuity was restored by means of an end-to-end direct anastomosis.

Takeuchi operation. A pedicled flap was developed in the anterior wall of the main PA, and the base toward the aorta. The aorta was opened obliquely, and an aortopulmonary window was created with a 4- or 4.5-mm puncher in the left wall of the aorta and at the root of the pedicled flap. Both holes were approximated together with a 6-0 absorbable suture. Then a coronary tunnel for the left coronary artery was created with the pedicled PA flap, which was approximated with the posterior wall of the main PA and along the nadir of the sinus of Valsalva and back to the site of the aortopulmonary window. The anterior defect in the main PA was covered with autopericardium.

Mitral annuloplasty. Simultaneous mitral annuloplasty at the anterolateral commissure was performed in all 24 patients with incompetence with polytetrafluoroethylene sutures. A pair of interrupted pledget-reinforced mattress stitches were placed through the annulus at the commissure (Fig 1). The commissure plication annuloplasty used here was similar to that described by Kay,4 Reed,5 and their associates. In addition, other plasty techniques were also used in 3 patients on the basis of the valve competence test after annuloplasty, as stated above, by injecting cold cardioplegic solution into the left ventricle (Fig 2). There was no significant difference in aortic crossclamp time between patients who had mitral annuloplasty and patients who did not (101 ± 29 vs 87 ± 21 minutes, P = .20). One patient without incompetence had an
associated closure of the atrial septal defect. No resection of the LV myocardium was performed in any patient.

**Statistical analysis.** Data computations were performed with the StatView 5.0 (SAS Institute Inc, Cary, NC) statistical program. The Mann-Whitney test was used to compare the crossclamp time for patients with and without mitral annuloplasty. The Fisher exact test was used to evaluate the relation of early age at operation, preoperative FSz, and preoperative mitral incompetence to mortality. The Wilcoxon signed-rank test for related data was used to analyze the differences between preoperative CTR or FSz values and postoperative values at different time points.

**Results**

**Survival.** There were 2 postoperative hospital deaths in infants who preoperatively had deteriorating LV function and severe mitral incompetence. One patient, with an FSz of –10.3, died 3 days after the operation because of ventricular fibrillation. The second patient (FSz of –7.2) died of low cardiac output, despite circulatory assistance of 7 hours. Early age at operation (<12 months), preoperative FSz (<–2), and preoperative mitral incompetence to mortality. The Wilcoxon signed-rank test for related data was used to analyze the differences between preoperative CTR or FSz values and postoperative values at different time points.

**Cardiac function.** Changes in the CTRs of operation survivors are shown in Fig 3. There were no significant differences between preoperative CTRs and those at the time of discharge (P = .35). However, at 1 and 5 years after surgical repair, the CTR decreased significantly (P = .005 and .003, respectively) compared with the preoperative ratios. FSz improved from –2.95 ± 3.90 at discharge compared with 0.56 ± 1.49 in the long-term period, and 15 of 16 patients showed FSz values above –2 (Fig 4).

**Mitral incompetence.** Preoperative mitral incompetence decreased (Fig 5) both at the time of discharge (none in 6, mild in 14, and moderate in 2 patients) and at the last follow-up (none in 10 and mild in 11 patients) in all but 1 operative survivor with mitral annuloplasty. One patient with infarction of both papillary muscles (Fig 2, C) had a mitral valve replacement 28 months after reimplantation (Fig 5). In the other 5 patients who had no incompetence or trivial incompetence preoperatively, that degree was unchanged at the last follow-up (data not shown).

**Reintervention.** Supravalvular pulmonary stenosis developed in 3 patients after the Takeuchi operation. One patient underwent surgical repair 5.7 years after the initial operation. Two patients underwent percutaneous transluminal angioplasty 5.3 and 5.5 years, respectively, after the initial operation. No patient had supravalvular pulmonary stenosis after direct aortic reimplantation. There was 1 patient who underwent mitral valve replacement, as described above.
Discussion

There has been a consensus that 2-coronary repair is an appropriate surgical approach for ALCAPA because there remains a risk of sudden death related to a persisting ischemia when simple ligation is performed. Our results also support this consensus. The overall mortality rate after the establishment of a 2-coronary system at our institution was 6.9%. In other series the overall mortality rates after 2-coronary repair were as follows: 0% for Neirotti and colleagues (n = 12), Alexi-Meskishvili and associates (n = 11), Turley and associates (n = 11), and Cochrane and coworkers (n = 21); 14% for Schwartz and colleagues (n = 42); 16% for Lambert and coworkers (n = 39); and 23% for Vouhé and associates (n = 22). The first 3 reports with no mortality had relatively small numbers of patients. Our good results in a large series have special importance because our series was performed under a consistent surgical policy with regard to mitral incompetence in patients with ALCAPA. The operative survivors in this study showed excellent long-term results even more than 10 years after operation. The CTR, as shown in Fig 3, showed definite improvement 5 years after operation. All 18 patients who survived more than 5 years postoperatively were clinically asymptomatic and required no cardiac medication. FSz was normalized in the late postoperative period (Fig 4). Several possible mechanisms for the recovery of cardiac function after operation have been advocated. A histologic study showed structural cellular adaptation to a chronic myocardial hypoperfusion. This may explain the delay in FSz recovery. “Hibernating” myocardium, which was the explanation given for persistent contractile dysfunction but which also preserved myocardial viability, might be an alternative mechanism. LV remodeling without myocardial cellular hypertrophy may also contribute to postoperative recovery.

There was no significant difference in mortality among the factors of early age at operation, preoperative FSz, and a higher degree of mitral incompetence in our study. These are in contrast to other investigators’ findings. The severity of the preoperative LV dysfunction has been reported to be a significant risk factor for mortality. In a recent study the degree of mitral incompetence was a risk factor, whereas the severity of LV dysfunction was not. However, to clarify factors that have influence on mortality, analysis of more cases is required, as through a multi-institutional study.

Mitral valve operations in patients with mitral incompetence associated with ALCAPA are controversial. Several studies have recommended mitral annuloplasty for patients with severe mitral incompetence. On the other hand, Vouhé and coworkers advocated that the mitral valve should not be interfered with when the initial operation is performed in infancy. In their patients who did not undergo valvuloplasty, however, there were 2 operative deaths and 1 survivor with severe incompetence unchanged 6 months after operation among 11 patients with ALCAPA and moderate or severe preoperative incompetence. The reason why the authors came to this conclusion was uncertain. Moreover, it was reported in the article by Cochrane and colleagues that without annuloplasty, 7 of 16 patients had no improvement in regurgitation and that significant regurgitation persisted in at least one third of patients in the long term (median follow-up period, 6.5 years). On the contrary, in our series there was no significant regurgitation remaining in the late period (Fig 5). Mitral incompetence is thought to result from dilatation of the mitral valve anulus with severe LV enlargement and from selective ischemia to the anterior papillary muscle. Ischemic elongation of the anterior papillary muscle and its chordae cause the prolapse in the region of anterolateral commissure. Kay-Reed type annuloplasty, which was simple and not so time consuming, was quite effective at reducing regurgitation. Therefore, we think that simultaneous mitral annuloplasty at the anterolateral commissure is a logical approach for supporting cardiac output during the critical postoperative period.
Establishing the 2-coronary system is the theoretical goal of surgery for ALCAPA. We believe that direct aortic reimplantation (direct coronary transfer) is the choice of treatment if technically possible. Coronary transfer is currently popular by experience with arterial switch operations in neonates. Some studies have demonstrated the growth of anastomoses between the aorta and the coronary artery if an adequate tissue of coronary button was taken. However, the Takeuchi procedure (intrapulmonary tunnel repair) would be an alternative in some situations where direct aortic reimplantation is not suitable. Those situations are as follows: (1) the anomalous orifice is located in the non-facing sinus (anterior pulmonary sinus) or in the left lateral position of the left pulmonary sinus or (2) the patient is older, between early childhood and late adolescence, with well-developed collaterals around the pulmonary sinuses.

Issues that have been reported after tunnel repair are supravalvular pulmonary stenosis, tunnel obstruction, and aortic regurgitation. In our study 3 patients had late postoperative pulmonary stenoses caused by shrinkage of the equine pericardium used for covering the defect of the PA in the earlier part of our series. Therefore, we have used autologous pericardium for this purpose since 1991 and have found no supravalvular pulmonary stenosis. No patients in our study had tunnel obstruction or aortic regurgitation. Two extrapulmonary baffling techniques with a transected pulmonary arterial wall have been reported from Japan. The use of autologous arterial wall tissue appears to have the advantage of allowing for subsequent growth of the reconstructed structure. Therefore, these new techniques might be useful or optional if a long-term patency were demonstrated in patients undergoing an extrapulmonary baffle operation.

In conclusion, these long-term results are promising and demonstrate that impaired LV function normalized in the late period (even if it was below normal immediately after operation) after the establishment of a 2-coronary artery system. On the basis of our experience at the Heart Institute of Japan, we recommend that the simultaneous mitral annuloplasty should be performed at the time of operation for patients who have varying degrees of mitral incompetence with ALCAPA.

We thank Dr Nakamura Tsuyoshi for his contribution to statistical analysis.

Received for publication Aug 15, 2000; revisions requested Sept 21, 2000; revisions received Oct 30, 2000; accepted for publication Nov 6, 2000.

REFERENCES


