EXCELLENT LONG-TERM FUNCTIONAL OUTCOME AFTER AN OPERATION FOR ANOMALOUS LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

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Objective: The aim of this study was to review the results of operations for anomalous left coronary artery from the pulmonary artery and the late outcome for exercise capacity, left ventricular function, and mitral regurgitation. **Methods:** Twenty-one patients underwent operations over an 18-year period (median age, 9 months; range, 6 weeks–26 years) with a median follow-up of 6.5 years (range, 2 months–18 years). In addition to clinical and echocardiographic follow-up, patients at our institution were also investigated with radionuclide scans ($n = 10$) and treadmill exercise testing ($n = 8$). **Results:** There were no operative or late deaths (0%; 95% confidence interval [CI], 0% and 16%). Five patients required support with a left ventricular assist device. Eighteen patients are currently in New York Heart Association class I, and 3 patients are mildly symptomatic. On nuclear gated scan at a mean of 6 years after the operation, the left ventricular ejection fraction was 64% (SD, 3%) at rest and increased to 74% (SD = 3%) on exercise (95% CI for the difference, 6%, 14%; $P = .001$). Treadmill endurance was normal for age (9.8–14.5 minutes) in those old enough to exercise. On echocardiography ($n = 18$), the current fractional shortening was 34% (SD, 4%) in the 15 patients with normal or only mildly abnormal ventricular septal motion. Three patients have undergone mitral valve operations. The left ventricular end-diastolic dimension fell from 48 mm (SD, 5.8 mm) before surgery to 35.1 mm (SD, 5.2 mm) at 1 year after the operation, and the fractional shortening increased over the first year from 19.6% (SD, 9.3%) to 32.8% (SD, 5.9%; both $P < .001$). **Conclusions:** Long-term clinical outcome and left ventricular function are good, despite severe left ventricular dysfunction at presentation. (J Thorac Cardiovasc Surg 1999;117:332–42)

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is an uncommon congenital defect that is often lethal if not diagnosed and treated.¹ The pathophysiologic condition depends on the pressure difference between the systemic and pulmonary arterial circulations and on the adequacy of col-

lateral vessels between the right and left coronary systems. The mechanism to explain the clinical presentation was proposed by Edwards,² who emphasized the dynamic nature of the pathophysiologic condition. Inadequate collateral flow results in left ventricular (LV) infarction, ranging from subendocardial to transmural in extent but often patchy in distribution, and causes ischemic damage to the papillary muscles, resulting in mitral regurgitation.

Several surgical procedures have been described, initially palliative, but more recently restoring a 2-coronary system, including various forms of bypass graft from the aorta,^{3,4} subclavian artery anastomosis,⁵ intrapulmonary bypass graft,⁶ intrapulmonary baffle,⁷ and direct coronary translocation to the aorta.^{8–10}

We reviewed our experience between 1980 and 1998 with this defect, the late clinical results, and the time-course of recovery of LV function. In addition, the con-

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Read at the Seventy-eighth Annual Meeting of The American Association for Thoracic Surgery, Boston, Mass, May 3–6, 1998.

Received for publication May 8, 1998; revisions requested June 5, 1998; revisions received Sept 16, 1998; accepted for publication Sept 16, 1998.

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0022-5223/99 \$8.00 + 0 12/6/94643

Fig 1. Preoperative aortogram. **A**, The figure demonstrates a dilated right coronary artery; no left coronary artery is seen. **B**, The figure shows late filling of the left coronary artery and passage of contrast into the MPA.

dition of all patients available for detailed review at our institution was investigated to assess global and regional LV function, mitral regurgitation, and exercise capacity.

Patients and methods

From January 1980 to March 1998, 21 patients underwent an operation for ALCAPA. The records were reviewed to document the clinical features, operative procedures, and the perioperative course.

All 10 patients living in the state of Victoria underwent clinical review at the hospital, and the cases were reinvestigated. The clinical status of the other patients was reviewed by contact with the treating cardiologists, and the reports of the most recent echocardiograms were obtained. Clinical follow-up was achieved on all patients.

Patient sample. Among the 21 infants and children, there were 13 female patients and 8 male patients. The median age at examination and operation was 9 months (range, 6 weeks–26 years); 13 of the 21 patients were less than 12 months of age.

Fourteen patients who experienced heart failure and severe LV impairment had a younger median age of 5 months (range, 6 weeks–22 months). Five patients had a murmur of mitral regurgitation and impaired LV function, without evidence of heart failure, at a median age of 2 years (range, 9

months–5 years). Two patients experienced chest pain at 11 and 26 years of age; the younger patient had undergone ligation of the left main coronary artery in infancy.

Mitral regurgitation was present in 19 patients at the time of diagnosis (severe in 5 patients, moderate in 10 patients, and mild in 4 patients). One other patient had undergone mitral valve replacement (23-mm mechanical valve) at 10 months of age, before the diagnosis of ALCAPA was made 15 months later. The anomalous coronary artery was not diagnosed until the persistently impaired LV function after the valve operation led to reinvestigation.

One infant who experienced severe heart failure, mitral regurgitation, and pericardial effusion on echocardiography was found to have a bloody effusion with an incipient rupture of the anterior LV wall because of transmural infarction. Extensive infarct resection and LV repair were performed with aortic implantation of the coronary artery.

Early in this experience in one other infant, the diagnosis of ALCAPA was missed until postmortem. She was examined at 7 weeks of age with severe heart failure, and a diagnosis of idiopathic dilated cardiomyopathy was made. She improved very little on medical therapy until death at 8 months of age. In all other patients, an operation was performed within hours to days of the diagnosis of the anomalous coronary artery. Seventeen of 21 patients had angiography before surgical repair (Fig 1). In the other patients, the diagnosis was clear on

Fig 2. Operative technique for aortic implantation. This figure demonstrates completion of the coronary artery transfer and patch repair of the pulmonary artery sinus. (From Buxton B, Frazier OH, Westaby S, editors. Ischemic heart disease: surgical management. London: Mosby-Wolfe, 1998. By permission of Mosby International, Ltd.)

Fig 2. Operative technique for aortic implantation. This figure demonstrates completion of the coronary artery transfer and patch repair of the pulmonary artery sinus. (From Buxton B, Frazier OH, Westaby S, editors. Ischemic heart disease: surgical management. London: Mosby-Wolfe, 1998. By permission of Mosby International, Ltd.)

echocardiography. No infant was refused an operation because of poor LV function.

Two children have undergone valve replacement (one before the recognition of the coronary lesion), and one other child underwent concomitant mitral valve repair because there was severe regurgitation and the child was almost 2 years of age.

Coronary artery anatomy. In 20 patients, the whole left main coronary artery was involved. In one infant, who experienced heart failure and mitral regurgitation, the circumflex coronary artery alone arose from the main pulmonary artery (MPA). The circumflex artery was subsequently shown to be "dominant" and supplied the posterior descending artery.

In 18 patients, the anomalous artery arose from the MPA, in 2 patients at the origin of the right pulmonary artery and in one patient from the mid-right pulmonary artery.

Surgical procedures. The 2 common forms of repair used were the intrapulmonary baffle (Takeuchi repair, $n = 12$) and direct aortic implantation ($n = 7$), with increasing use of aortic implantation in the latter half of the series.

Direct implantation was performed by mobilization of a large button of pulmonary artery wall around the anomalous coronary ostium, often removing the whole of the sinus, and creation of a trap-door flap in the aorta. The technique of coronary prolongation described by Sese and Imoto¹¹ was used in 2 patients, creating an inferior aortic wall flap and a

Table I. Comparison of the investigational group with patients followed up at other institutions

	Investigational group ($n = 10$)	Others ($n = 11$)	P value
Age at surgery (mo; median)	4.7	10.5	.14
Sex (M:F)	3:7	5:6	>.20
Fractional shortening % at presentation (median)	15	23	.03
LVAD support	3	2	>.20
Fractional shortening on most recent study (%; median)	33	35	>.20
Follow-up time (y; median)	6.5	5.2	>.20

superior pulmonary arterial flap. The pulmonary sinus was repaired with autologous pericardium (Fig 2).

One 11-year-old patient, who had undergone coronary ligation in infancy, was examined again in 1983 with possible angina; a saphenous vein bypass graft was the choice of therapy at the time because the ostium had been obliterated.

The infant with the anomalous circumflex coronary artery underwent ligation of the circumflex artery by a left thoracotomy, in the belief that the myocardial insult from a circumflex artery lesion was likely to be limited and because the patient was quite small (2.3 kg). However, the infant required nasogastric-tube feeding for many months and continued medical therapy for heart failure with mitral regurgitation.

Investigational subgroup. A subgroup of 10 infants and children were recently reviewed in our institution and investigated further with transthoracic echocardiography, a radionuclide scan, and a treadmill exercise test. The median age at review was 7.2 years (range, 13 months–16.9 years), with 7 female patients and 3 male patients. The median time from the operation was 6.5 years (range, 9 months–16.7 years). The procedures included intrapulmonary baffle repair ($n = 5$), aortic implantation ($n = 4$), and ligation of an anomalous circumflex artery ($n = 1$). Three patients had required postoperative support with an LV assist device (LVAD).

Nine of the 10 patients were completely asymptomatic, in New York Heart Association (NYHA) class I, although the child who had undergone circumflex ligation had persisting exertional dyspnea. Two patients received angiotensin-converting enzyme inhibitors. One patient had undergone mitral valve replacement subsequent to the primary repair; 6 of the other 9 patients had residual murmurs of mitral regurgitation. The investigational group is compared with the other patients in Table I. There is no difference between the groups except for the fractional shortening percent on echocardiography before surgery, which was significantly worse in the investigational group. This excludes any selection bias between the 2 groups.

Echocardiographic assessment. The most recent echocardiographic results were available for 18 patients (median age at echocardiography, 7.2 years; interquartile range, 4.2–12.5

years; median body surface area, 0.93 m²; interquartile range, 0.68–1.36 m²). The median postoperative time to study was 5 years (interquartile range, 15 months–9.3 years).

Transthoracic 2-dimensional, M-mode, and color Doppler echocardiography was performed on all patients. Ten patients were studied at our institution, and the studies were reviewed by one of the authors (D.M.C.). For the other 8 patients, who lived outside of Victoria, the latest echocardiography result was obtained from their cardiologists. The findings assessed were the LV end-diastolic dimension (LVEDD) with respect to body surface area,¹² LV shortening fraction corrected for age,¹³ ventricular septal motion, and the presence and degree of mitral regurgitation. The shortening fraction was not taken into account in patients with flat or paradoxical ventricular septal motion. Mitral regurgitation was graded by color flow Doppler mapping as absent, trivial to mild, moderate, or severe according to a combination of the width of the regurgitant jet at the mitral valve orifice and the extent of the jet into the left atrium. In patients who had undergone an intrapulmonary baffle repair, the presence of any baffle leak and/or increased peak systolic flow velocity in the MPA was noted.

To assess the time course of change in LV fractional shortening and LVEDD after the time of operation, all recorded measurements were grouped into 5 time periods (preoperative, 1 week after the operation, 1 to 6 months, 9 to 18 months, and more than 2 years from the operation).

Exercise testing. A treadmill test was performed in 8 children, all of whom were over 5 years of age and cooperative. Two children were too young to perform the test. Resting 12-lead electrocardiography was performed, followed by a Bruce exercise protocol.¹⁴ Continuous electrocardiographic monitoring was performed throughout the exercise test and until 6 minutes after the completion of the test. The blood pressure change was measured at peak exercise, and the duration of exercise was recorded. Exercise duration and peak heart rates were compared with pediatric normal values.¹⁵ Cardiopulmonary testing was not performed.

Radionuclide cardiac studies. Gated nuclear scans were performed in all 10 patients, and exercise was possible in 7 patients. The first-pass right ventricular ejection fraction was measured in the right anterior oblique projection. The LV ejection fraction (LVEF) was measured in the left anterior oblique projection at rest and during graded semisupine bicycle exercise.

Statistical methods. The confidence limits for death were determined by the binomial exact technique; the 1-sided 97.5% confidence limit was used because there were no deaths. Freedom from reoperation was analyzed by the Kaplan-Meier method. The radionuclide ejection fractions before and after exercise were compared by paired *t* test. Longitudinal changes over time in LV fractional shortening and LVEDD were compared by linear regression, with standard errors calculated from the information sandwich formula for repeated measurements within individual patients.¹⁶ Data were not available on each patient in every time period, but all available data were included in the analysis. This involves an assumption that data were missing at random, which was

checked by repeating the analysis for fully observed patients only to ensure that the conclusions did not change.

Statistical calculations were performed with Stata Statistical Software (release 5.0; Stata Corp, College Station, Texas). Results are expressed as the mean and standard deviation for normally distributed data and as the median, range, and interquartile range for skewed data.

Results

Early and late clinical status. There were no perioperative or 30-day deaths (0%; 95% CI, 0% and 16%). There have been no late deaths over a median follow-up period of 6.5 years (range, 2 months–18 years) with a total of 145 patient-years of follow-up.

Currently, 18 patients have normal clinical exercise ability (NYHA class I). Three patients are mildly symptomatic (NYHA class II) with tachypnea on exertion as the main symptom. One of these children, now 2 years old, who underwent circumflex ligation at 6 weeks of age, has moderate to severe mitral regurgitation and requires continued medical therapy, although LV function was nearly normal on a contrast left ventriculogram and the radionuclide LVEF was 59% at rest. The second infant, now aged 2 years, who underwent coronary implantation at 10 months of age, has occlusion of the implanted left main coronary artery on angiography with collateral flow from the right coronary artery (“ligation equivalent”) and also requires continued medical therapy.

Three patients are being treated with angiotensin-converting enzyme inhibitors (including the 2 patients just described); 2 patients with mechanical valves require warfarin therapy, and 2 patients are being treated with aspirin. No patient requires diuretics or digoxin therapy.

Since 1989, 5 of 12 patients undergoing an operation have received LVAD support when there has been significant difficulty in weaning from cardiopulmonary bypass. The mean preoperative fractional shortening in the 5 patients was 12.4% (SD, 1.7%). Support was required for a median of 3 days (range, 2–4 days), and there has usually been significant clinical improvement in cardiac function observed during this period, as measured by inotropic drug requirement. Two patients in this group had neurologic deficits, with full recovery in one patient. No other patient in the series had a neurologic deficit.

Two patients underwent subsequent procedures, one procedure for mitral valve replacement at 19 months after the initial operation and the other procedure for the repair of pulmonary artery stenosis at 7 years after a Takeuchi repair. This provides an 84% freedom from reoperation at 10 years (95% CI, 47% and 96%; Fig 3).

Fig 3. Kaplan-Meier analysis for freedom from reoperation after repair of anomalous left coronary artery, with the 95% confidence limits included.

Exercise capacity. In the 8 patients who underwent testing, the mean endurance was 12 minutes (SD, 1.8 minutes; range, 9.8-14.5 minutes). All performed above the 10th percentile for their age, and 5 of the 8 performed above the 50th percentile. The blood pressure rise was normal in 7 patients; the other patient did not have measurement at peak exercise but had a normal blood pressure response during exercise for the radionuclide scan. All patients reached 90% or more of maximum predicted heart rate, with a peak heart rate of 193 beats/min (SD, 5 beats/min; range, 185-201 beats/min).

There was no chest pain or significant arrhythmia. However, one exercise test demonstrated 2 mm of horizontal ST depression, commencing at 10 minutes of exercise. This patient has a small fistula from the intrapulmonary baffle to the MPA.

Late LV function. On radionuclide examination ($n = 10$), the mean LVEF at rest was 64% (SD, 8%; range, 50%-73%; normal laboratory reference range, 50%-70%). In the 7 patients who were old enough to perform semisupine exercise, the LVEF rose significantly from 66% (SD, 9%) to 76% (SD, 8%; 95% CI for the difference, 6% and 14%; $P = .001$), with the heart rate increasing from 78 (SD, 8) to 148 (SD, 15) beats/min. There were no wall motion defects observed on radionuclide study, except in the infant who had required infarct resection and LV repair and who had reduced anteroseptal wall motion, but this patient's global LVEF was within the normal range at 64%. The mean first-pass right ventricular ejection fraction was 65% (SD, 6%; range, 57%-76%; laboratory normal reference range, 45%-65%).

The most recent echocardiographic examination ($n = 18$) included 2 patients without dual coronary revascu-

larization. Three patients had significantly abnormal ventricular septal motion. In these 3 patients, the LV posterior wall contractility was described as normal in 2 of the patients and reasonable in the patient with an occluded left main coronary repair. In the other 15 patients, the mean LV shortening fraction was 34% (SD, 4%; range, 30%-40%). However, mildly flattened ventricular septal motion in 5 of these patients may have reduced the calculated shortening fraction. The shortening fraction was normal for age in 9 of the remaining 10 patients and mildly reduced (30% at 13 months of age) in 1 patient with at least moderate mitral regurgitation. LVEDD was normal for body surface area in 12 patients and increased in 5 patients, 3 of whom had at least moderate mitral regurgitation.

Increased echogenicity of the papillary muscles and LV endocardium was noted in 10 of 18 patients, consistent with old ischemic injury and fibrosis.

Two patients, after Takeuchi repair, have baffle leaks into the MPA. One of these patients had ST segment changes on exercise testing and achieved 14.5 minutes on the treadmill. Four other patients who had baffle procedures, including the patient who underwent late repair of the MPA, have a mildly increased peak systolic flow velocity (1.9-2.4 m/s) in the MPA on echocardiography. One patient has mild pulmonary valve stenosis.

Time course of LV recovery. The mean preoperative fractional shortening ($n = 15$) was 19.6% (SD, 9.3%). At 1 week after the operation ($n = 10$), it remained similar at 20.8% (SD, 9.0%; $P > .20$). There was a significant improvement by 1 to 6 months after the operation ($n = 18$) to 28.4% (SD, 10.3%; mean difference, 8.8%; 95% CI for the difference, 1.6% and 16.0%; $P = .02$)

Fig 4. Time-related changes in LV fractional shortening percentage, before the operation (n = 15), at 1 week (n = 10), at 1 to 6 months (n = 18), at 1 year (n = 14), and beyond 2 years (n = 34). * $P < .05$; ** $P < .01$; *** $P < .001$.

and marked recovery by 1 year after repair (n = 14) to 32.8% (SD, 5.9%; mean difference, 13.3%; 95% CI for the difference, 7.0% and 20%; $P < .001$). At late follow-up (all measurements >2 years from the operation; n = 34) the recovery was maintained ($P < .001$, compared with preoperative level; Fig 4). The proportion with an abnormal shortening fraction fell from 80% before the operation to 9% at 1 year.

The changes in LVEDD were analyzed in a similar fashion to fractional shortening (Fig 5). The LVEDD before the operation (n = 12) was markedly enlarged for a predominantly infant population at 48.3 mm (SD, 5.8 mm). It fell rapidly within the first week of the operation (n = 10) to 37.3 mm (SD, 6.2 mm; mean difference, 11 mm; 95% CI for the difference, 7 mm and 15 mm; $P < .001$) and was significantly less at all subsequent time points ($P < .001$, at 1 to 6 months [n = 16]; at 1 year [n = 13]; and $P = .01$ at late follow-up [n = 35] compared with preoperation). The maximal fall occurred at 1 year after the operation to 35.1 mm (SD, 5.2 mm; mean difference, 13.2 mm; 95% CI for the difference, 8 mm and 18 mm). The modest increase between 1 year and late follow-up (range, 2-16 years) reflects the normal cardiac growth in these children.

Late mitral valve regurgitation. Among the 21 patients, 3 patients have undergone mitral valve surgery. Of the 18 patients with available recent echocardiographic results, 5 patients have at least moderate mitral regurgitation (including the 2 patients without revascularization); 9 patients have trivial to mild regurgitation; 2 patients have no mitral regurgitation, and 2 patients have a mitral prosthesis. The severity of regurgitation immediately before the operation and on the latest fol-

low-up echocardiogram were compared. Nine of 16 (56%) patients with a dual coronary repair had a reduction in regurgitation of 1 grade or more at late follow-up, although 44% had minimal or no improvement.

Discussion

This study demonstrates that excellent early and late survival can be obtained with current operations. The late clinical status is generally good; with all except 3 patients in NYHA class I, all children tested were able to achieve a normal level of exercise and there was good recovery of LV function. However, the important residual problem in at least one third of patients has been significant mitral regurgitation.

Surgical technique. Our approach, like that of other groups, has evolved over the time of this experience, aided by increasing confidence in coronary transfer from the arterial switch procedure. We currently regard direct aortic implantation of the anomalous artery as the preferred technique, and there are several methods described for lengthening of the coronary pedicle.^{11,17,18} However, our late results suggest no difference in the outcome between aortic implantation and the intrapulmonary baffle. By the use of a large pericardial patch to the anterior wall of the MPA with the baffle repair, the incidence of pulmonary artery stenosis has been low compared with that in some other series.¹⁹

LV function. The late results indicate nearly complete recovery of LV function, despite severe impairment at initial examination, confirming previous work.¹⁹⁻²¹ This recovery begins in the first few days after the operation, demonstrated by the rapid recovery in infants requiring LVAD support, and is usually com-

Fig 5. Time-related changes in LVEDD, before surgery (n = 12), at 1 week (n = 10), at 1 to 6 months (n = 16), at 1 year (n = 13), and beyond 2 years (n = 35). * $P < .05$, ** $P < .01$, *** $P < .001$.

plete by 1 year after the operation. This capacity for rapid recovery is an excellent example of myocardial hibernation.²¹

However, mild, but persisting, ventricular enlargement is seen in several patients, sometimes but not always explained by mitral regurgitation; and abnormalities of ventricular septal motion are also common, indicating residual effects of the ischemic insult. This is consistent with pathologic studies demonstrating widespread but patchy endocardial fibrosis despite revascularization.²³

This study demonstrates that the first sign of the recovery in LV function is a rapid reduction in cardiac size, measured as LVEDD, within 1 week of the operation, even though the fractional shortening has not changed at that stage. Subsequently, the fractional shortening progressively recovers over 1 year, similar to the time course reported by others.^{19,20}

Mitral regurgitation is an almost universal feature at presentation, because of a combination of papillary muscle ischemia or infarction and subsequent fibrosis, LV free wall dyskinesis, and LV dilation. Our policy is to not repair the valve at the time of coronary repair, because this increases the cardiac ischemic time, and improvement usually occurs after recovery of LV function. This improvement is slower than the changes seen in cardiac size and contractility. Furthermore, adult cardiac experience has demonstrated the difficulty often found in achieving major improvement for "ischemic mitral regurgitation" by valve repair.

However, significant mitral regurgitation persists in

at least one third of patients, despite good recovery of LV function. This usually reflects the presence of irreversible papillary muscle infarction and scarring. Therefore, in an older child undergoing a primary operation for ALCAPA with established mitral regurgitation, concomitant valve repair is warranted.

Study limitations. The presence of mitral regurgitation limits the accurate assessment of LV function, in particular on the radionuclide examination, because it overestimates the LVEF percent. However, the normal response to exercise on the radionuclide study and the normal exercise duration in these children suggest that the level of cardiac function is quite satisfactory.

The grading of mitral regurgitation by Doppler echocardiography is often qualitative and at best semi-quantitative. The degree of mitral regurgitation at earlier times in this study was determined from echocardiograms reported by a number of observers, although the most recent echocardiograms in the investigational subgroup (n = 10) were performed by one observer (D.M.C.). Given the time period of the study (18 years), developments in 2-dimensional and Doppler echocardiography over this period, and the geographic dispersion of the patients, the evaluation of mitral regurgitation may have varied because of different grading techniques and observers.

Abnormal ventricular septal motion may cause difficulty with interpretation of the LV fractional shortening, and we acknowledge that, in the assessment of time-related changes, we have taken no account of this abnormality. However, the presence of abnormal septal

motion would reduce the measured fractional shortening and therefore should not alter the broad conclusion.

For the study of time-related changes in LV function, our analysis attempted to account for the problem of "missing data." Fifteen patients had satisfactory preoperative measurements, but the number and timing of the postoperative studies varied between patients, with a total of 91 studies in the group at different times. The presence of missing data was greater in the earlier patients and in patients referred from distant centers. The analysis assumes that the data are missing at random. The alternative explanation would be that there is a bias and that the sicker patients with poorer postoperative function may have had closer follow-up and more studies. However, although this may change the magnitude of the improvement observed, it would not alter the basic conclusion that even infants with severely impaired LV function will usually show a dramatic recovery of LV function.

Experience with a single coronary circulation.

Coronary ligation was performed in one patient with isolated origin of the circumflex artery. In retrospect, this infant may have done better with revascularization. The circumflex territory initially appeared quite small on angiography, but subsequent angiography at 2 years of age suggests that it is a dominant artery supplying the whole of the inferior and lateral LV walls and that the initial appearance may have simply reflected the poor collateralization, which resulted in early onset of symptoms. The very early onset of symptoms at 6 weeks of age with heart failure and mitral regurgitation also suggests that the circumflex artery was serving a significant portion of the myocardium. The continuing symptoms and mitral regurgitation in the child with an occluded repair ("ligation equivalent") add further evidence for the benefit of revascularization over ligation.

LVAD support. This has become an important adjunct to pediatric cardiac operations.²⁴ Infants with ALCAPA provide one of the ideal groups for LVAD use, because there is predominant LV dysfunction, often transiently exacerbated by the operation ("myocardial stunning") and subsequent rapid improvement in LV function. This favorable experience in patients with ALCAPA has also been reported by other centers.²⁰

Place of heart transplantation. Some authors have performed heart transplantation for infants with this lesion and severely impaired LV function.^{25,26} Our experience, and that of others,²⁰ would suggest that there is little indication for transplantation as the primary surgical treatment. Even when LVAD support is required, late cardiac function has been good.

We thank the cardiologists in other states of Australia who provided follow-up information: Dr Jim Ramsay and Dr Luigi d'Orsogna (Perth), Dr Brodie Knight and Dr Malcolm Richardson (Adelaide), and Dr Chris Whight (Brisbane).

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Discussion

Dr Yasuharu Imai (*Shinjuku-ku, Japan*). I would like to congratulate Dr Cochrane and his colleagues for their excellent results. There were no operative deaths and no late deaths in their series of 21 patients, and the importance of LVAD support in 5 high-risk infants is emphasized.

Our surgical experience in ALCAPA comprised 29 cases in the past 17 years, and we also intended to establish a 2-coronary system in all cases using direct aortic implantation in 18 cases and Takeuchi operation in 11 cases. I find that the best advantage of the Takeuchi procedure is coronary rerouting in situ, when there is no redundancy in the left coronary artery in infancy. Therefore, if there is any doubt, we prefer to do Takeuchi procedure in infants.

A major difference arises in the concomitant mitral repair. In our series of 29 cases, 23 infants had a significant mitral regurgitation, ranging from moderate to severe, so we did mitral annuloplasty at the anterior commissure in 23 patients concomitantly with the repair of the coronary artery. As you know, the mechanism of mitral regurgitation in this subset was infarction or ischemic elongation of the anterior papillary muscle and its chordae, which caused the prolapse in the

region of anterolateral commissure. These pathological changes in supportive structure did not seem to improve significantly with time. Standard Kay-Reed-type annuloplasty at the anterior commissure was quite effective to reduce regurgitation even in the immediate postoperative period. Therefore in our series there is not much of the significant regurgitation remaining in the postoperative period.

Regarding LV function, I agree completely with Dr Cochrane. Within several years, LV function became normalized, in our case 62% in average, ranging from 55% to 67%. LV end-diastolic volume decreased also markedly an average of 126% of normal, ranging from 101% to 145%.

What is your criterion of concomitant mitral repair, because only 2 patients had mitral valve replacement or repair at the time of operation?

What is your practical management of LVAD including anesthetic agent, vasodilator, phenoxybenzamine, optimal blood flow, ultrafiltration unit, if you have optimal blood pressure to be maintained, and criteria for weaning?

Were there any differences in the ischemic times in direct aortic implantation and Takeuchi operation in your series?

Dr Cochrane. You mentioned first that you prefer to use the intrapulmonary baffle, and I think we would have no major disagreement with that. That obviously has been the major technique of repair, although more recently we have moved to aortic implantation. However, I would emphasize that there is no difference in the long-term results between those 2 groups. I think that, for a surgeon who is less experienced and is concerned about aortic implantation, the intrapulmonary baffle is a very appropriate operation. However, because we have had increasing experience with the arterial switch and with mobilization of coronaries, we have become more comfortable with the reimplantation.

In terms of the crossclamp times, the ischemic times, they are rather longer with aortic implantation, and I think that is one factor that has to be considered, particularly if LV function is very poor. The mean crossclamp time in the Takeuchi intrapulmonary baffle procedure is about 42 minutes and for aortic implantation about 55 minutes.

With regard to LVAD, there were 5 patients. Their median age was 5 months, so it was the younger infants. They had particularly poor preoperative fractional shortening, mean of 12%. Support was required for a median of 3 days, ranging from 2 to 4 days. Two patients had neurologic complications with good resolution in one patient, but persisting hemiparesis in one other patient.

We looked at the risk factors for LVAD use, looking at age, degree of mitral regurgitation, fractional shortening, and diastolic dimension. Although none of these are significant, because the numbers are inadequate, I would emphasize that there does appear to be a trend toward increased risk of LVAD use in patients with larger ventricles and poorer fractional shortening. The Z score in the 5 patients with LVAD was 9.6, nine standard deviations away from the mean, compared with 5.7 in the patients without LVAD with a high odds ratio but very wide confidence limits because the numbers are small and similarly, for fractional shortening, 12% in the patients

with LVAD compared with 19%. Again I would emphasize that the numbers are inadequate, but there are some trends.

With regard to the LVAD management, I think I can address all of those issues. We do not routinely use dilators, but we use them as appropriate according to the mean perfusion pressure. We would aim to run a mean arterial pressure around 40 to 50 in most infants on LV assist support. We would run full flow, which for us means 150 mL/kg per minute. In general, recovery of function, as I have shown, is fairly quick. We would expect to see ejection by the second or certainly the third day after the operation. We would then wean to a low flow in the intensive care unit on a small dose of dopamine and assess function clinically and on echocardiography, using transesophageal echocardiography if necessary, and would then, if the findings appeared satisfactory, return to the operating theater for decannulation.

Finally, in terms of the place of mitral valve operations, we have not performed mitral valve operations initially, and we believe that the most important thing is revascularization to allow recovery of myocardium. The reasons that we do not perform repair in the infant group follow: First, in two thirds, it is not necessary. It means a longer crossclamp time in a damaged heart in those patients. We believe that the mitral operation is certainly easier technically at a later time in childhood when annuloplasty rings can be used along with other procedures, which are significantly more difficult in the infant group where one is restricted more or less to a commissural annuloplasty. We know from adult experience that ischemic mitral regurgitation can be difficult to repair, certainly to get complete repair. However, I would agree that in an older child, above 2 years of age, if there is significant mitral regurgitation, that mitral repair at the time of operation is very reasonable and that we probably would do that.

Mr Magdi Yacoub (*London, England*). In your series, you have used a variety of techniques for revascularization. Have you looked at anatomic or functional characterization of either the proximal or distal coronary circulation associated with each technique? We have used a method of anatomic correction of the anomaly by threading the button behind the pulmonary artery, implanting it from inside the aorta in the middle of the left coronary sinus, and we think that has functional implications. Have you looked at either anatomic or functional characteristics of the coronary circulation in the different techniques?

Dr Cochrane. I think I can partly answer the question. Quite a number of these patients have had angiography at a later time. And anatomically, in all cases, there appears to be a wide open ostium and no obvious difference.

In terms of function, we have said that LV function is normal; but in terms of coronary endothelial function, I do not think I can make any comment if that is the aspect you are referring to.

Mr Yacoub. And the site of implantation, do you think that is important whether it is in the sinus or not?

Dr Cochrane. I do not think that the site of implantation into the aorta is particularly important. I think, in most cases,

that it really depends on what appears to be anatomically the most appropriate site.

Dr Vladimir Alexi-Meskishvili (*Berlin, Germany*). In our group in Berlin, we had the opportunity in the last 7 years to treat 23 patients with this anomaly. Among them were 18 children and infants; the youngest was 3 weeks old. In all patients we prefer to perform direct implantation of the coronary artery. But contrary to your group, we prefer to correct mitral incompetence during the implantation because we think that correcting the mitral incompetence provides better cardiac output after the operation. None of our patients needed mitral valve surgery later on during follow-up.

Do you consider, in the future, reconstruction of the mitral valve in patients with severe mitral incompetence and enlarged left atrium, which is sometimes seen by patients with this anomaly?

We, too, successfully used the LVAD in our patients in 3 cases, and I am interested in your indication for this kind of assist. Do you use long reperfusion after the unsuccessful attempt to wean patients from the heart-lung bypass or how do you make the decision to use an assist device in your patients with this anomaly?

Dr Cochrane. With regard to the place of mitral valve repair, you report excellent results from early concomitant mitral valve repair, and it is difficult to comment because we have had a different philosophy. I can only emphasize that two thirds of patients have done well with significant improvement and would not have needed valve repair; however, I recognize that there are different philosophies and methods of management.

In terms of the indications for LV assist support, clearly there have been a number of surgeons involved over a period of time, so that we do not have rigid criteria. In most cases, there would have been a significant period of reperfusion after removal of the crossclamp, and there might well have been several attempts at weaning from bypass. The usual indications would be inability to wean, having had 1 or 2 attempts to wean from bypass, and excessively high inotropic requirement, by which I mean that we were needing a very high dose of dopamine or that epinephrine had been necessary to institute. We believe that, rather than causing further damage to these hearts with excessive doses of epinephrine, it is much better for recovery of the heart to place the patient on LV assist. Usually it is necessary for a median of only 3 days.

Dr Zohair Y. Al-Halees (*Riyadh, Saudi Arabia*). We also have had gratifying results in 10 patients with the same anomaly that we corrected at our institution. In all of the patients, a 2-coronary artery system was established. Though our follow-up is not as long as yours and is a maximum of about 9 years, we observed improvement in all of the patients. The mean ejection fraction improved from 35% to 57% (fractional shortening, 19%–31%). However, we noticed that the ventricles that were poorer to start with showed less recovery. We also analyzed the electrocardiograms in all patients. We observed ischemic changes preoperatively in the form of abnormal Q waves in leads I, aVL, V4–V6, with ST segment depression of more than 0.1 mm. There was evi-

dence of left axis deviation for age in 5 patients and LV hypertrophy in 8 patients. These changes normalized postoperatively, particularly the signs of ischemia.

Did you notice, what we observed, that poorer ventricles tend to show less recovery? Have you looked at the electrocardiograms of the patients and noticed similar changes to the ones we described?

Dr Cochrane. With regard to the electrocardiogram, yes, I think that the changes are similar and that they have been widely reported before. The electrocardiogram improves quite markedly, and in the long term there may be only minimal residual changes. So yes, I would certainly agree with those findings, but they have been reported extensively before.

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