

Long-Term Results of Repair of Anomalous Origin of the Left Coronary Artery From the Pulmonary Artery

Rüdiger Lange, MD, PhD, Manfred Vogt, MD, PhD, Jürgen Hörer, MD, Julie Cleuziou, MD, Andrea Menzel, MD, Klaus Holper, MD, PhD, John Hess, MD, PhD, and Christian Schreiber, MD, PhD

Departments of Cardiovascular Surgery, and Paediatric Cardiology and Congenital Heart Defects, German Heart Center Munich, Technical University Munich, Munich, Germany

Background. Upon initial repair, most patients with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) present with severely impaired ventricular function and mitral regurgitation. In this study, both parameters were investigated at long-term in patients in whom either coronary transfer or subclavian artery anastomosis was applied.

Methods. Records of 56 patients with ALCAPA, operated between 1977 and 2002, were reviewed retrospectively. Patients were divided into two groups: subclavian artery anastomosis group ($n = 25$), and coronary transfer group ($n = 31$).

Results. Concomitant mitral valve repair was performed in one patient. Thirty-day mortality was 14.3% (subclavian artery anastomosis group, $n = 3$; coronary transfer group, $n = 5$). Preoperative age ($p = 0.007$) and left ventricular ejection fraction (LVEF) less than 0.35 ($p = 0.020$) were predictive for 30-day mortality, whereas preoperative mitral regurgitation, surgical technique,

and cardiopulmonary bypass time were not. For both groups, LVEF ($p = 0.006$) and mitral regurgitation ($p = 0.013$) were improved at the time of hospital discharge. Mean follow-up was 11.0 ± 7.2 years. Survival of hospital survivors at 20 years was $94.8 \pm 3.6\%$. At final follow-up, 95.5% of the patients presented with a LVEF greater than 0.50, and 84.1% with a mitral regurgitation less than grade 2. Mitral valve replacement was performed in three patients.

Conclusions. In the majority of patients with ALCAPA, both ventricular function and mitral valve regurgitation normalize over time. Concomitant mitral reconstruction may not be required upon initial repair. Coronary transfer is the technique of choice today. However, subclavian artery anastomosis may remain an option in selected cases.

(Ann Thorac Surg 2007;83:1463–71)

© 2007 by The Society of Thoracic Surgeons

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital anomaly, first described by Brooks in 1882 [1]. It usually presents as an isolated lesion and accounts for 1 of 300,000 live births (0.25% to 0.5%). In 1933 the clinical spectrum of ALCAPA was described by Bland, White, and Garland [2], therefore ALCAPA in Europe is also referred to as the Bland-White-Garland syndrome.

Different surgical approaches have been suggested, including ligation of the anomalous left coronary artery [3], various forms of bypass grafts from the aorta [4, 5], subclavian-left coronary artery anastomosis [6], transpulmonary baffling or Takeuchi procedure [7], and direct reimplantation of the coronary artery into the aorta [8]. Currently, immediate surgical correction upon diagnosis with the aim of restoring a two-coronary circulation system is considered the standard treatment for patients with ALCAPA.

Accepted for publication Nov 1, 2006.

Address correspondence to Dr Hörer, German Heart Center Munich, Dept. of Cardiovascular Surgery, Lazarettstr. 36, Munich, D-80636 Germany; e-mail: hoerer@dhm.mhn.de.

Improvement of left ventricular function after successful reestablishment of a two-coronary system was uniformly documented [9, 10]. However, mitral valve reconstruction upon the initial repair has been discussed controversially. While some authors recommended concomitant mitral annuloplasty with the initial repair [11, 12], others clearly demonstrated that mitral valve function generally improves after ventricular remodeling without any intervention [13, 14].

Only a few long-term studies on large collectives are available, and different techniques for repair have not yet been evaluated with respect to the fate of ventricular and mitral valve functions. Hence, we compared the long-term results after either coronary transfer or subclavian anastomosis based on our single-center experience.

Patients and Methods

Patients

All patients in whom either coronary transfer ($n = 31$) or subclavian artery anastomosis ($n = 25$) was applied for ALCAPA repair between 1977 and 2002 were included

Table 1. Preoperative Variables of 56 Patients With ALCAPA Treated With Subclavian Artery Anastomosis (n = 25), or Coronary Transfer (n = 31)

Characteristics	Overall Group (n = 6)	Subclavian Artery Anastomosis (n = 25)	Coronary Transfer (n = 31)	p ^a
Gender: male, n (%)	36 (64.3)	15 (60.0)	21 (67.7)	0.586
Operation before 1988	28 (50.0)	17 (68.0)	11 (34.5)	0.031
Mean age at operation [months]	16.2 ± 29.3	10.4 ± 13.3	20.9 ± 37.2	0.153
Mean weight at operation [kg]	7.9 ± 5.8	7.3 ± 3.6	8.5 ± 7.2	0.452
Mean height at operation [cm]	72 ± 21	69.1 ± 14.6	74.3 ± 24.4	0.358
Preoperative MR > I°, n (%)	32 (57.1)	11 (44.0)	21 (67.7)	0.105
Preoperative EF > 0.35, n (%)	35 (62.5)	13 (52.0)	22 (71.0)	0.173
Cardiopulmonary bypass, n (%)	52 (92.9)	21 (84.0)	31 (100.0)	0.034
Mean cardiopulmonary bypass time [min]	99 ± 50	62 ± 33	128 ± 41	<0.001
Aortic cross-clamp, n (%)	28 (50.0)	7 (28.0)	21 (67.7)	0.007
Mean ischemic time [min]	31 ± 37	12 ± 21	47 ± 40	<0.001
Total circulatory arrest, n (%)	10 (17.9)	5 (20.0)	5 (16.1)	0.738
Mean circulatory arrest time [min]	6 ± 15	6 ± 14	6 ± 15	0.972
Postoperative MR > I°, n (%) ^b	15 (31.3)	5 (22.7)	10 (38.5)	0.351

^a p = level of significance for the comparison between the subclavia anastomosis and the coronary transfer group. the initial operation, excluding the eight early deaths.

^b Within hospital admission after

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery; EF = ejection fraction; MR = mitral regurgitation.

into a retrospective follow-up study. Patients in whom a Takeuchi procedure (n = 3) or a left mammary artery graft (n = 3) were performed were excluded, as well as one patient with a concomitant Norwood procedure. This study was approved by the ethics committee of the Technical University Munich. Informed consent was obtained from each patient, or parent.

Data Collection

The clinical records were reviewed to document the clinical features, operative procedures, and the perioperative course. The latest echocardiographic examination was reviewed by one of the authors. Ventricular function was classified as normal or mildly impaired (ejection fraction [EF] > 0.50), moderately impaired (EF 0.35 to 0.50), and severely impaired (EF < 0.35). Ventricular function was assessed in M-mode in the parasternal long and short axis view. Mitral regurgitation (MR) was evaluated with Doppler echocardiography (0 = no MR; 1 = mild MR; 2 = moderate MR; 3 = severe MR; 4 = severest MR).

The composite endpoint was defined as left ventricular EF (LVEF) less than 0.50, MR grade 2 or more, or death after hospital discharge after the initial operation.

Statistics

Descriptive data for continuous variables are presented as means ± standard deviation or as medians with ranges; categoric variables are presented as relative frequencies. The Fischer exact test or Mc Newar test was performed to detect significant differences between groups. For comparison of continuous variables between two groups, the *t* test was used (two-tailed tests were used for all analyses). The probability of freedom from

events was estimated according to the Kaplan-Meier method. Freedom-from-events curves were compared by means of the log-rank test. Values of *p* less than 0.05 were considered as statistically significant. According to Concato and colleagues [15] multivariate analysis could not be performed because the ratio of events per variable was too small. Analyses were performed with SPSS 12.0.2 for Windows (SPSS Inc, Chicago, IL).

Results

Preoperative Data

Thirty-six male, and 20 female patients fulfilled the inclusion criteria. The mean age of both groups at time of repair was 16.2 ± 29.3 months (range, 13 days to 12 years). Two thirds of the patients presented with severely impaired left ventricular function and one third with more than mild mitral regurgitation. There were no significant differences in preoperative hemodynamic and demographic parameters between the subclavian artery anastomosis group and the coronary transfer group, except that significantly more patients who were operated on before 1988 received a subclavian artery anastomosis compared with patients who were operated on thereafter (*p* = 0.031; Table 1). Patients presenting with an EF below 0.35 were significantly younger (mean age, 9.3 ± 23.8 years) compared with patients presenting with an EF above 0.35 (mean age, 27.8 ± 34.3 years; *p* = 0.038). The distributions of patients according to LVEF and mitral valve regurgitation are depicted in Figures 1 and 2.

In the subclavian artery anastomosis group, the origin of the ALCAPA (aspect from the pulmonary trunk) was as follows: 9 posterolateral, 10 lateral, 4 posterior, and 2

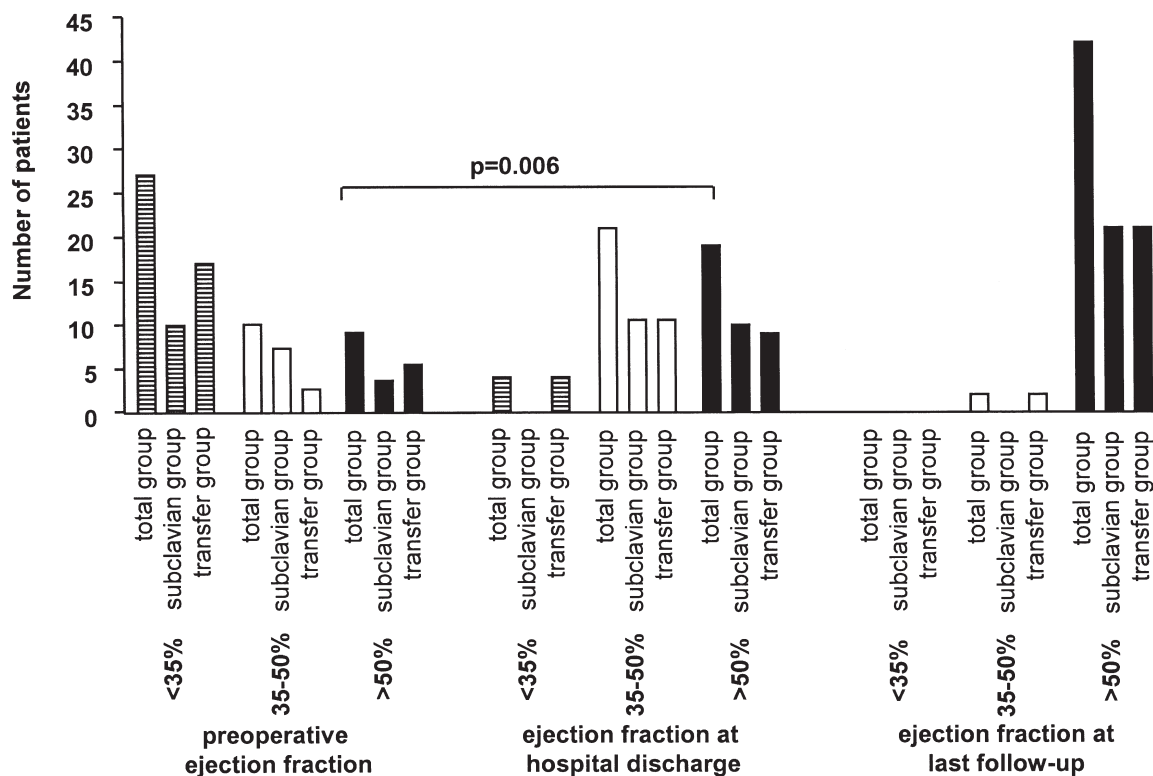


Fig 1. Distribution of patients according to left ventricular ejection fraction prior to anomalous origin of the left coronary artery from the pulmonary artery repair, at hospital discharge, and at last follow-up, stratified by the operative technique.

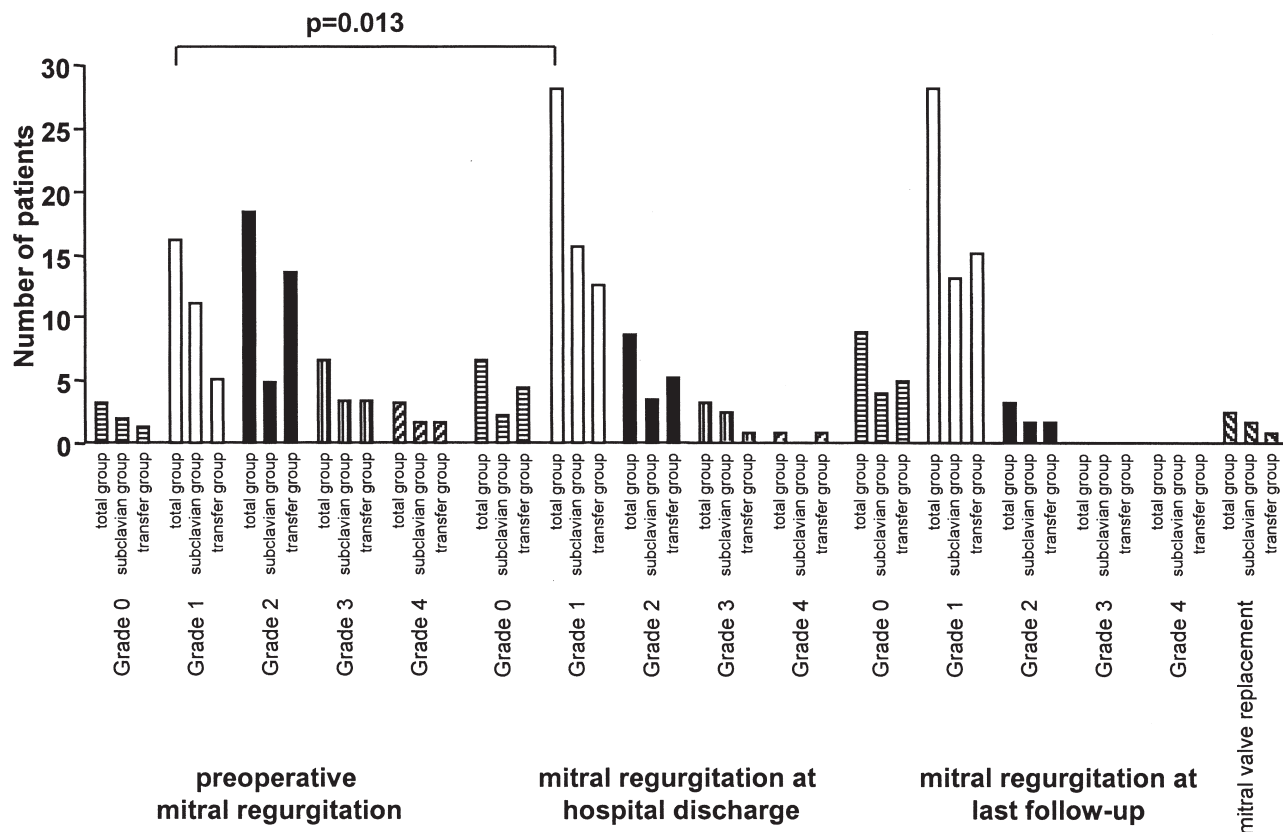


Fig 2. Distribution of patients according to mitral valve regurgitation prior to anomalous origin of the left coronary artery from the pulmonary artery repair, at hospital discharge, and at last follow-up, stratified by the operative technique.

unspecified, respectively. In the coronary transfer group, there were 9 posterolateral, 2 lateral, 15 posterior, 1 from the left pulmonary artery, 1 with a single-trunk anomalous origin of both coronary arteries, and 3 unspecified, respectively.

Surgical Technique

The subclavian-left coronary artery anastomosis was performed in an end-to-end fashion after excision of the coronary button from the pulmonary artery. Coronary transfer was performed by mobilization of the button, and creation of a trap-door flap in the aorta. Techniques of coronary elongation [16, 17], such as described by Sese and Imoto [18], were also used, creating an inferior wall flap and a superior pulmonary artery flap. The pulmonary artery was closed directly, or reconstructed with glutaraldehyde-fixed autologous pericardium or Gore-Tex patch material (Gore-Tex, Inc, Flagstaff, AZ), respectively.

Simultaneous mitral annuloplasty was performed in one patient, in whom the morphology of the mitral valvar apparatus was pathologic (distinct prolapse of the anterior leaflet). The patient was four-months old at the time of operation, and was treated with a Whooley annuloplasty and additional shortening of anterior leaflet chordae.

Operations were performed on cardiopulmonary bypass in 52 patients (93%). The aorta was clamped in 28 patients (50%). Crystalloid cardioplegia was administered simultaneously in both great vessels after cross-clamping and after snaring of both pulmonary arteries. In the subclavian artery anastomosis group, cardiopulmonary bypass time ($p = 0.034$) and cardiac arrest time ($p < 0.001$) were shorter than in the coronary transfer group (Table 1). Sixteen percent of the patients in the subclavian artery anastomosis group were operated without cardiopulmonary bypass.

Postoperative Data

In both groups, LVEF ($p = 0.006$) and MR ($p = 0.013$) improved from preoperative to the time of hospital

discharge (Figs 1; 2). There was no significant difference in the fate of left ventricular function and MR between the subclavian artery anastomosis group and the coronary transfer group (Table 1).

Early Mortality

Thirty-day mortality was 14.3% for both groups, with three deaths in the subclavian artery anastomosis group, and five deaths in the coronary transfer group, respectively. There was no mortality in the last 13 years (17 patients operated). Preoperative age ($p = 0.007$) and LVEF less than 0.35 ($p = 0.020$) were predictive for 30-day mortality (Table 2). Preoperative MR, surgical technique, cardiopulmonary bypass time, and cardiac arrest time did not influence 30-day mortality.

Follow-Up

Final follow-up could be obtained from 46 patients. Follow-up was complete for 95.8% of hospital survivors, with a maximum follow-up time of 23.8 years. Mean follow-up of the patients after the subclavian to coronary artery anastomosis was 14.5 ± 5.5 years (complete follow-up), and mean follow-up of the patients after the coronary transfer technique was 8.7 ± 6.9 years (two patients lost to follow-up), and thus significantly different ($p = 0.003$).

Late mortality was 3.3% (two patients). One patient (coronary transfer group) with an extremely rare morphology, a single-trunk anomalous origin of both coronary arteries, died four months after coronary transfer during a reoperation. One patient (subclavian artery anastomosis group) died eight years after initial repair due to malignant arrhythmias. This patient died three weeks after her last outpatient visit. She was clinically in heart failure due to severe MR and therefore had ventricular extrasystole with couplets and triplets (on 24-hour Holter monitoring). On last echo, there were no signs for myocardial failure (EF 0.65). Thus, it seems to be obvious that she had ventricular tachycardia as the most

Table 2. Predictors for Early Death in 56 Patients With ALCAPA Treated With Subclavian Artery Anastomosis ($n = 25$), or Coronary Transfer ($n = 31$)

Characteristics	Early Deaths ($n = 8$)	Early Survivors ($n = 48$)	p^a
Gender: male, n (%)	7 (87.5)	29 (60.4)	0.236
Operation before 1988	5 (62.5)	23 (47.9)	0.705
Mean age at operation [months]	5.1 ± 3.1	18.1 ± 31.3	0.007
Mean weight at operation [kg]	5.1 ± 1.0	8.4 ± 6.1	0.001
Mean height at operation [cm]	63 ± 3	74 ± 22	0.002
Preoperative MR $> I^\circ$ n (%)	3 (37.5)	29 (60.4)	0.268
Preoperative EF < 0.35 , n (%)	8 (100.0)	27 (56.3)	0.020
Subclavian artery anastomosis	3 (37.5)	22 (45.8)	0.720
Cardiopulmonary bypass, n (%)	7 (87.5)	45 (93.8)	0.470
Mean cardiopulmonary bypass time [min]	130 ± 74	94 ± 44	0.057
aortic crossclamp, n (%)	3 (37.5)	25 (52.1)	0.705
Mean ischemic time [min]	24 ± 38	32 ± 37	0.531

^a p = level of significance for the comparison between the early deaths and early survivors (30 days after initial operation).

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery; EF = ejection fraction; MR = mitral regurgitation.

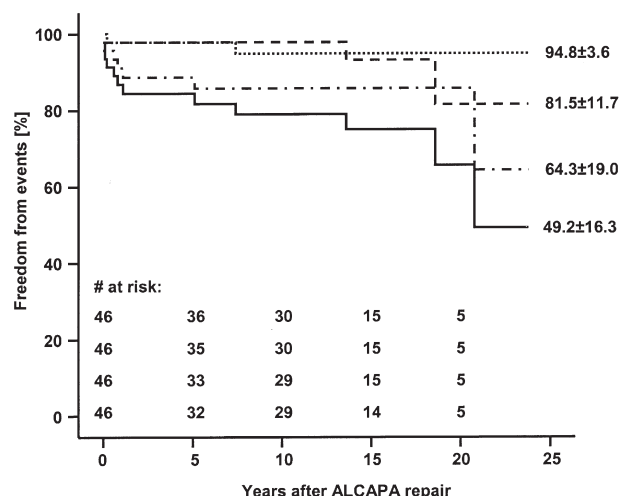


Fig 3. Freedom from left ventricular ejection fraction less than 0.50, mitral regurgitation greater than grade 1, death after hospital discharge, and composite endpoint in 46 patients referred for ALCAPA repair (Kaplan-Meier survival plot). (··· = late death; --- = ventricular dysfunction; - · - = mitral regurgitation; — = composite endpoint; ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery.)

obvious reason for sudden cardiac death. Estimated survival of hospital survivors at 20 years was $94.8 \pm 3.6\%$.

Three patients presented during follow-up with an EF less than 0.50, with no evidence of impaired coronary circulation upon angiography. At final follow-up one of these patients presented in New York Heart Association class I (NYHA I). Two patients are symptomatic, one is in NYHA II, with tachypnea on exertion, and one suffers from severely impaired ventricular function, remaining currently in NYHA II-III. Accordingly, freedom from more than mildly impaired ventricular function at 20 years was $81.5 \pm 11.7\%$. At final follow-up, 95.5% of the patients presented with a LVEF greater than 0.50 (Fig 1).

Seven patients exhibited moderate or severe MR during the follow-up period. Three of these underwent mitral valve replacement: One patient (age, 14 months) underwent two attempts for reduction of mitral valve insufficiency one year postoperatively due to a hugely dilated annulus and significant prolapse of the anterior leaflet, and finally had valve replacement (St Jude, size 21 mm; St Jude Medical, Inc, Minneapolis, MN) the same year. One patient (age, 17 months) had mitral valve replacement (size 23 mm; St. Jude Medical, Inc) after 13 months. One patient (age, 4 years) had aortic valve replacement with a homograft and mitral valve replacement (size 25 mm; CarboMedics, Inc, Austin, TX) after one month, and subsequent aortic valve replacement (size 16 mm; ATS Medical, Inc, Minneapolis, MN) after five years. Freedom from MR greater than grade 1 at 20 years was $85.7 \pm 5.4\%$ (Fig 3). Three additional patients were reoperated. One patient (age, 12 years) had aortic valve replacement (size 23 mm; CarboMedics) five years after initial repair. Pulmonary artery augmentation was performed in two patients (age, 7 and 4 years) after three

and five years, of which both had had previous subclavian anastomoses. Both patients had primary closure of the pulmonary artery. Freedom from reoperation at 20 years was $81.0 \pm 6.2\%$, with no difference between the subclavian artery anastomosis group and the coronary transfer group.

Hence, 12 patients reached the composite endpoint of LVEF less than 0.50, MR greater than grade 1, or death after hospital discharge after the initial operation. Freedom from composite endpoint at 10 and 20 years was 78.9 ± 6.3 , and 65.6 ± 10.8 , respectively (Fig 3). Preoperative weight ($p = 0.042$), preoperative height ($p = 0.038$), preoperative MR greater than grade 1 ($p = 0.028$), cardiopulmonary bypass time ($p = 0.012$), and postoperative MR greater than grade 1 ($p < 0.001$) were predictive for the composite endpoint, whereas other demographic parameters and the surgical technique had no influence (Table 3).

Presently, 13 patients had cardiac catheterization over time; four patients had scintigraphy, and three patients had positron emission tomography investigation. Angiographic examples for the coronary transfer and the subclavian anastomosis are depicted in Figures 4 and 5, respectively. Currently, four patients are on angiotensin-converting enzyme inhibitors, two of whom are on a combination with digitoxin. Two patients are on β -blocker therapy and two on amiodarone.

Table 3. Predictors for Freedom From Composite Endpoint of Death, Mitral Regurgitation, or Impaired Ventricular Function in 44 Patients with ALCAPA Treated With Subclavian Artery Anastomosis ($n = 22$) or Coronary Transfer ($n = 24$)

Characteristics	No./Events	p^a
Gender, male	27/7	0.901
Age at operation < 5.8 months (median)	23/7	0.200
Weight at operation < 6.2 kg (median)	23/8	0.042
Height at operation < 64 cm (median)	23/8	0.038
Preoperative MR > I°	27/10	0.028
Preoperative EF < 35%	26/6	0.818
Subclavian artery anastomosis	22/5	0.162
Cardiopulmonary bypass	43/11	0.828
Cardiopulmonary bypass time > 90 min (median)	23/8	0.012
Cardiac arrest	23/6	0.579
Ischemic time > 7 min (median)	23/6	0.579
Postoperative MR > I°	13/8	<0.001
No improvement in MR	24/6	0.745
Postoperative EF < 0.35	4/2	0.074
Postoperative EF < 0.50	27/8	0.571
No improvement in EF	18/6	0.435

^a p = level of significance between groups.

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery; EF = ejection fraction; MR = mitral regurgitation.

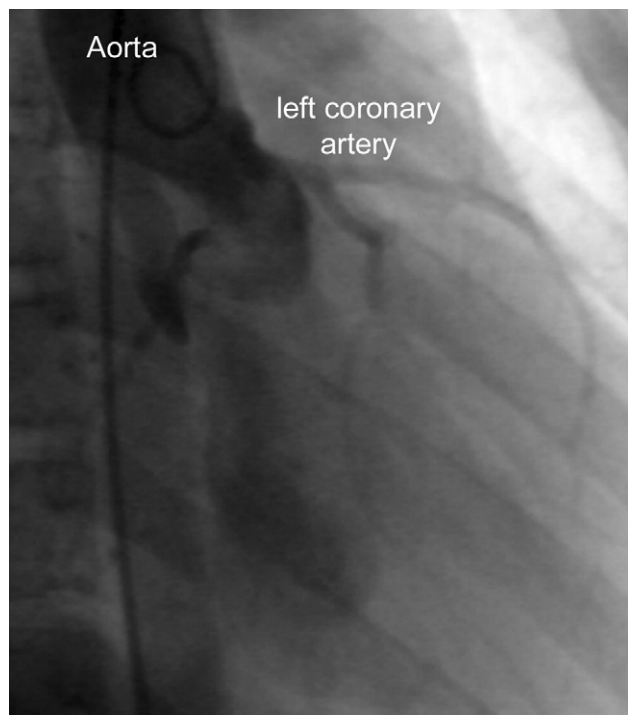


Fig 4. Coronary angiogram of a 20-year-old patient, 20 years after coronary transfer.

Comment

Initial attempts for the repair of ALCAPA included the creation of an aortopulmonary anastomosis, thereby increasing pulmonary artery bloodflow and, hence, left coronary artery oxygen saturation [19]. A more physiologic approach was achieved in 1953 by Mustard, creating an end-to-end anastomosis between the left common carotid artery and the ALCAPA [20], and Cooley and colleagues [5] reported in 1966 the creation of a two-coronary system using a saphenous vein graft. Two years later, Meyer and colleagues [6] presented the case of a patient who had a successful anastomosis of the left subclavian artery to the origin of the ALCAPA. It was not before 1974 that Neches and colleagues [21] first described the technique of direct implantation of the anomalous left coronary artery into the aorta, by transferring it with a button of pulmonary artery wall. In 1979, Takeuchi and colleagues [7] proposed a solution for cases, where at the time direct implantation did not seem feasible, creating a tunnel between the coronary ostium in the pulmonary artery and the aorta.

In the present study, the subclavian artery anastomosis and coronary transfer technique were investigated. We thought to ascertain if both techniques were equally effective in the long-term recovery of left ventricular and mitral valve function.

Most patients with ALCAPA present with symptoms of severely impaired left ventricular function and mitral valve regurgitation. Left ventricular damage resulting from myocardial ischemia in patients with ALCAPA is associated with a range of pathologic findings. These

include endocardial and subendocardial fibrosis, damage to the papillary muscles, patchy myocardial necrosis, and dilatation of the ventricle [22]. As a consequence, mitral incompetence frequently develops [23, 24]. In the present study, 62.5% of the patients exhibited a preoperative EF below 0.35, and 37.5% showed moderate to severe MR.

Various risk factors for operative survival have been identified in the treatment of ALCAPA, including the preoperative impairment of LV function [25–27] and young age at operation [27]. In the present study, both parameters were also found to be significant predictors for 30-day mortality. Because younger patients presented with significantly lower LVEF, we believe that small size and low age at the operation are markers for earlier clinical manifestation of myocardial malperfusion. This might be explained by the original anatomy: patients with good collateralization might be asymptomatic for a long time and become evident only by a cardiac murmur. Patients with little collateralization become symptomatic early in life, with cardiac failure due to ischemic heart lesions.

The severity of preoperative MR has also been identified as a risk factor for survival after surgery [28], although this is not supported unanimously [8, 29, 30]. Looking at our patients, it could not be identified as a risk factor for the 30-day mortality. Based on our experience, we have developed the policy not to address MR by repair or replacement at the initial operation. In ALCAPA, even severe mitral insufficiency has been re-

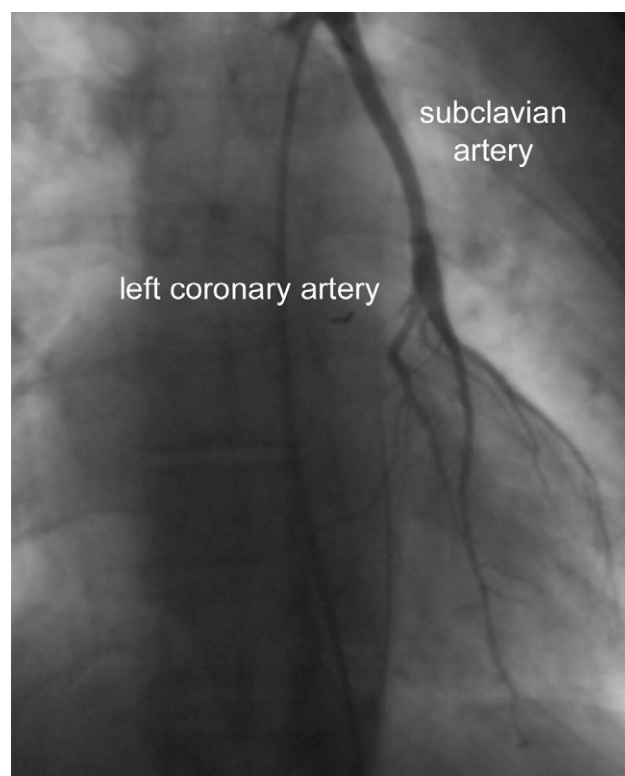


Fig 5. Coronary angiogram of a 28-year-old patient, 25 years after subclavian artery anastomosis.

ported to fully regress after reperfusion alone in the majority of cases [14, 31]. In the present study, mitral valve regurgitation had already improved significantly by the time of hospital discharge when compared with the preoperative finding. Furthermore, freedom from MR greater than grade 1 at 20 years was $85.7 \pm 5.4\%$. Thus, the primary goal in surgery should be coronary reperfusion and myocardial salvage. Presumably, the added ischemic time to perform mitral valve procedures in the setting of severely compromised ventricular function, is potentially more deleterious than helpful [13, 14]. Furthermore, the results from surgical treatment of ischemic MR in adults are relatively disappointing [32], and “over-correction” of mitral annulus diameter is not an option in children. Other techniques for correction of ischemic regurgitation, such as Whooler annuloplasty, artificial chordae, chordal shortening, and others may not be applicable in newborns and infants. Thus, the policy of not approaching the mitral valve upon the initial operation, but rather wait for ventricular remodeling to ensue, is also advocated by many others [8, 11, 16, 33]. Early postoperative mild-to-moderate mitral insufficiency is usually acceptable [8, 11, 16] and, after improvement of left ventricular function, persistently symptomatic or hemodynamically significant MR may be treated at a later time [8, 13, 34, 35]. Interestingly, our data showed that mitral function recovers completely in the early postoperative period in most patients. In contrast, ventricular function recovered incompletely during hospital stay, but improved further in the long term. An opposing finding has been reported by Azakie and colleagues [36]. The authors found that improvements in MR lagged behind normalization of EF and left ventricular dilatation. In our experience, the first phase of ventricular remodeling after ischemic dilatation of the left ventricle is a reduction in left ventricular diameters that can be measured on M-mode. When the left ventricle becomes smaller ventricular systolic and diastolic function improves. These geometrical and functional changes (smaller mitral valve orifice, less stroke volume, and better cardiac output) might contribute to an improvement in MR.

Some authors [12], however, recommend routine mitral valve repair at the time of coronary artery revascularization on the grounds that early postoperative cardiac output is improved and operative mortality reduced. It remains speculative whether their excellent results would have been achieved without associated mitral valve procedures. Others advocate mitral annuloplasty [11, 29, 37] or mitral valve replacement [37, 38] only in the presence of severe MR. In our patients, the mitral valve was addressed during the initial repair in only one patient, who presented with associated mitral valve pathology not related to ischemia. However, in the long term, three patients needed additional treatment for persistent severe mitral valve regurgitation, and the mitral valve eventually was replaced in all cases. Hence, postoperative mitral incompetence was identified as a risk factor for reintervention, but not for death.

Mechanical assist devices or extracorporeal membrane oxygenation were not used in any of our patients. The question of the need for mechanical circular support in patients undergoing ALCAPA repair is addressed in a series of 16 patients corrected by direct aortic reimplantation [33]. These authors state that ventricular support systems may only be necessary in very few selected cases if ventricular function does not improve early postoperatively, despite adequate coronary revascularization. Using repeated antegrade crystalloid cardioplegia with simultaneous administration to both great vessels, combined with appropriate postoperative inotropic support, may allow for favorable results in the majority of the cases, without the need of mechanical circulatory support. Since 1992, no patient ($n = 17$) has died at our institution applying this strategy. Because of excellent results worldwide, we also do not think that concomitant resection of left ventricular muscle during initial surgery, as has been proposed in the past, is still justified today [8, 17, 26].

At present, coronary reimplantation is the technique of choice for repair of ALCAPA and may be performed in almost every case [16]. The present study confirms excellent perioperative and long-term results with this technique, which has also been reported by others [14, 28]. Our approach toward coronary transfer, like that of other groups, has evolved over time, aided by increasing confidence in the arterial switch procedure. Several methods have been described for lengthening of the coronary pedicle, such as creating an inferior wall flap and a superior pulmonary artery flap facilitating coronary transfer in complicated cases [16–18]. Very rarely, the Takeuchi tunnel technique may also still be appropriate, if the coronary ostium lies extremely lateral and so close to the pulmonary valve commissure that excision may not generate sufficient arterial wall to support coronary transfer. In the present study group, the subclavian artery anastomosis was preferentially performed in patients with a lateral position of the ostium of the left coronary artery (10 of 12 patients). The surgeons might have seen a problem in translocating the coronary artery to the aorta. However, with the growing experience in coronary transfer by the arterial switch operation, presently, coronary transfer is considered to be possible in almost all of the cases, at least in neonates and small children.

Half of the patients of the present study population were operated before 1988. Two-thirds of them received a subclavian artery anastomosis, whereas only two-fifths of the patients who were operated thereafter received a subclavian artery anastomosis. At the time when surgeons were not yet as acquainted with the coronary reimplantation technique from the arterial switch procedure, the subclavian artery anastomosis seemed to be a safe alternative, especially in small children. Cardiopulmonary bypass time was shorter and, according to our experience, 16% of the patients could even be operated on without cardiopulmonary bypass. The present investigation revealed no difference between the two revascularization techniques for any parameter studied. To our

knowledge, only one other report exists about the long-term results (average 10-year follow-up) of the subclavian artery to coronary artery anastomosis in seven infants with ALCAPA [27]. Similar to the findings of the present investigation, the authors report on good exercise tolerance and normalization of ventricular function. Hence, although the subclavian artery to coronary artery anastomosis is no longer performed, this technique yielded the same results as the now generally accepted coronary transfer technique. Our current series with this technique is, to our knowledge, the largest reported to date with the longest follow-up, showing that it was, in the past, a valid option for repair of ALCAPA; especially as it could be performed without cardiopulmonary bypass or temporary aortic occlusion.

In conclusion, our findings show that patients with ALCAPA surviving the perioperative period have an excellent prognosis for both immediate improvement of mitral valve function and functional recovery of the left ventricle in the long term. Because preoperative impairment of ventricular function turned out as a risk factor for operative mortality in our analyses, we suggest that patients with ALCAPA should undergo surgery as soon as the diagnosis is confirmed, in order to salvage myocardial tissue, regardless of the finding that age at repair was revealed also as a risk factor. In most cases, the mitral valve can safely be approached at a later time. Left subclavian artery to coronary artery anastomosis and direct implantation of the left coronary artery were equally effective surgical techniques.

References

1. Brooks H. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effects of this anomaly in producing cirroid dilation of the vessel. *J Anat* 1882;20:26-9.
2. Bland EF, White PD, Garland J. Congenital anomalies of coronary arteries. Report of unusual case associated with cardiac hypertrophy. *Am Heart J* 1933;8:787-801.
3. Sabiston DC Jr, Neill CA, Taussig HB. The direction of blood flow in anomalous left coronary artery arising from the pulmonary artery. *Circulation* 1960;22:591-7.
4. Chaitman BR, Bourassa MG, Lesperance J, Grondin P. Anomalous left coronary artery from pulmonary artery. An eight year angiographic follow-up after saphenous vein bypass graft. *Circulation* 1975;51:552-5.
5. Cooley DA, Hallman GL, Bloodwell RD. Definitive surgical treatment of anomalous origin of left coronary artery from pulmonary artery: indications and results. *J Thorac Cardiovasc Surg* 1966;52:798-808.
6. Meyer BW, Stefanik G, Stiles QR, Lindesmith GG, Jones JC. A method of definitive surgical treatment of anomalous origin of left coronary artery. A case report. *J Thorac Cardiovasc Surg* 1968;56:104-7.
7. Takeuchi S, Imamura H, Katsumoto K, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg* 1979;78:7-11.
8. Vouhe PR, Baillot-Vernant F, Trinquet F, et al. Anomalous left coronary artery from the pulmonary artery in infants. Which operation? When? *J Thorac Cardiovasc Surg* 1987;94:192-9.
9. Arciniegas E, Farooki ZQ, Hakimi M, Green EW. Management of anomalous left coronary artery from the pulmonary artery. *Circulation* 1980;62:1180-9.
10. Levitsky S, van der Horst RL, Hastreiter AR, Fisher EA. Anomalous left coronary artery in the infant: recovery of ventricular function following early direct aortic implantation. *J Thorac Cardiovasc Surg* 1980;79:598-602.
11. Alexi-Meskishvili V, Hetzer R, Weng Y, et al. Anomalous origin of the left coronary artery from the pulmonary artery. Early results with direct aortic reimplantation. *J Thorac Cardiovasc Surg* 1994;108:354-62.
12. Isomatsu Y, Imai Y, Shin'oka T, Aoki M, Iwata Y. Surgical intervention for anomalous origin of the left coronary artery from the pulmonary artery: the Tokyo experience. *J Thorac Cardiovasc Surg* 2001;121:792-7.
13. Huddleston CB, Balzer DT, Mendeloff EN. Repair of anomalous left main coronary artery arising from the pulmonary artery in infants: long-term impact on the mitral valve. *Ann Thorac Surg* 2001;71:1985-9.
14. Cochrane AD, Coleman DM, Davis AM, Brizard CP, Wolfe R, Karl TR. Excellent long-term functional outcome after an operation for anomalous left coronary artery from the pulmonary artery. *J Thorac Cardiovasc Surg* 1999;117:332-42.
15. Concato J, Peduzzi P, Holford TR, Feinstein AR. Importance of events per independent variable in proportional hazards analysis. I. Background, goals, and general strategy. *J Clin Epidemiol* 1995;48:1495-501.
16. Turley K, Szarnicki RJ, Flachsbarth KD, Richter RC, Popper RW, Tarnoff H. Aortic implantation is possible in all cases of anomalous origin of the left coronary artery from the pulmonary artery. *Ann Thorac Surg* 1995;60:84-9.
17. Vigneswaran WT, Campbell DN, Pappas G, Wiggins JW, Wolfe RW, Clarke DR. Evolution of the management of anomalous left coronary artery: a new surgical approach. *Ann Thorac Surg* 1989;48:560-4.
18. Sese A, Imoto Y. New technique in the transfer of an anomalously originated left coronary artery to the aorta. *Ann Thorac Surg* 1992;53:527-9.
19. Potts WJ, Kittle CF, Diehl AM, Heilbrunn A. Anomalous left coronary artery arising from the pulmonary artery. *J Pediatr* 1955;47:196-203.
20. Ravitch MM, Welch KJ, Benson CD, Aberdeen E, Randolph JG. *Pediatric Surgery*. 1st ed. Chicago: Year Book Medical Publishers; 1982.
21. Neches WH, Mathews RA, Park SC, et al. Anomalous origin of the left coronary artery from the pulmonary artery. A new method of surgical repair. *Circulation* 1974;50:582-7.
22. Smith A, Arnold R, Anderson RH, et al. Anomalous origin of the left coronary artery from the pulmonary trunk. Anatomic findings in relation to pathophysiology and surgical repair. *J Thorac Cardiovasc Surg* 1989;98:16-24.
23. Noren GR, Raghbi G, Moller JH, Amplatz K, Adams P Jr, Edwards JE. Anomalous origin of the left coronary artery from the pulmonary trunk with special reference to the occurrence of mitral insufficiency. *Circulation* 1964;30:171-8.
24. Burchell HB, Brown AL. Anomalous origin of the left coronary artery masquerading as mitral insufficiency. *Am Heart J* 1962;63:388-93.
25. Elhendy A, Zoet-Nugteren S, Cornel JH, et al. Functional assessment of ALCAPA syndrome by dobutamine stress thallium-201 SPECT and echocardiography. *J Nucl Med* 1996;37:748-51.
26. Singh TP, Di Carli MF, Sullivan NM, Leonen MF, Morrow WR. Myocardial flow reserve in long-term survivors of repair of anomalous left coronary artery from pulmonary artery. *J Am Coll Cardiol* 1998;31:437-43.
27. Kesler KA, Pennington DG, Nouri S, et al. Left subclavian-left coronary artery anastomosis for anomalous origin of the left coronary artery. Long-term follow-up. *J Thorac Cardiovasc Surg* 1989;98:25-9.
28. Schwartz ML, Jonas RA, Colan SD. Anomalous origin of left coronary artery from pulmonary artery: recovery of left ventricular function after dual coronary repair. *J Am Coll Cardiol* 1997;30:547-53.

29. Lambert V, Touchot A, Losay J, et al. Midterm results after surgical repair of the anomalous origin of the coronary artery. *Circulation* 1996;94:II38–43.
30. Sauer U, Stern H, Meisner H, Buhlmeyer K, Sebening F. Risk factors for perioperative mortality in children with anomalous origin of the left coronary artery from the pulmonary artery. *J Thorac Cardiovasc Surg* 1992;104:696–705.
31. Jin Z, Berger F, Uhlemann F, et al. Improvement in left ventricular dysfunction after aortic reimplantation in 11 consecutive paediatric patients with anomalous origin of the left coronary artery from the pulmonary artery. Early results of a serial echocardiographic follow-up. *Eur Heart J* 1994;15:1044–9.
32. Galloway AC, Grossi EA, Spencer FC, Colvin SB. Operative therapy for mitral insufficiency from coronary artery disease. *Semin Thorac Cardiovasc Surg* 1995;7:227–32.
33. Backer CL, Hillman N, Dodge-Khatami A, Mavroudis C. Anomalous origin of the left coronary artery from the pulmonary artery: successful surgical strategy without assist devices. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2000;3:165–72.
34. Backer CL, Stout MJ, Zales VR, et al. Anomalous origin of the left coronary artery. A twenty-year review of surgical management. *J Thorac Cardiovasc Surg* 1992;103:1049–58.
35. Stern H, Sauer U, Locher D, et al. Left ventricular function assessed with echocardiography and myocardial perfusion assessed with scintigraphy under dipyridamole stress in pediatric patients after repair for anomalous origin of the left coronary artery from the pulmonary artery. *J Thorac Cardiovasc Surg* 1993;106:723–32.
36. Azakie A, Russell JL, McCrindle BW, et al. Anatomic repair of anomalous left coronary artery from the pulmonary artery by aortic reimplantation: early survival, patterns of ventricular recovery and late outcome. *Ann Thorac Surg* 2003;75:1535–41.
37. Yam MC, Menahem S. Mitral valve replacement for severe mitral regurgitation in infants with anomalous left coronary artery from the pulmonary artery. *Pediatr Cardiol* 1996;17:271–4.
38. Bunton R, Jonas RA, Lang P, Rein AJ, Castaneda AR. Anomalous origin of left coronary artery from pulmonary artery. Ligation versus establishment of a two coronary artery system. *J Thorac Cardiovasc Surg* 1987;93:103–8.

INVITED COMMENTARY

Since the initial reports on the surgical management of anomalous left coronary artery from the pulmonary artery more than 40 years ago, several different surgical techniques have been used. More recently, and borrowing from experience with the arterial switch operation, restoration of a two-coronary circulation by direct reimplantation of the anomalous coronary artery seems to be the preferred surgical approach by most surgeons. Due to the low incidence of this anomaly and the usual practice of embracing one surgical approach, comparative studies looking at recovery of ventricular function, mitral insufficiency, and the need for reintervention afforded by different techniques are limited and lack meaningful follow-up.

The article by Lange and colleagues [1] provides an opportunity to characterize and compare the mid-term outcome of two surgical techniques in a sizable cohort of patients with well-documented assessments of ventricular and mitral function during follow up. The selective use of subclavia flap or coronary reimplantation approach allowed the inclusion of both techniques throughout the duration of the study period, eliminating some of the bias of noncontemporary comparisons.

With similar rates of early and late mortality, as well as recovery of ventricular function and mitral insufficiency it is not surprising that Lange and colleagues [1] have not

abandoned one technique in favor of the other, but rather embraced the selective application of each of them. The ability to perform the subclavian flap repair with a shorter period of myocardial ischemia, cardiopulmonary bypass, and in selected cases no cardiopulmonary bypass at all seems very attractive, and this may provide an edge over the more conventional direct translocation technique.

The study by Lange and colleagues [1] provides possibly not only the longest follow-up on the subclavian artery anastomosis, but also a good argument to consider this technique in an era when direct coronary translocation seems to be the best approach.

Christian Pizarro, MD

*Nemours Cardiac Center
Alfred I. duPont Hospital for Children
1600 Rockland Rd
Wilmington, DE 19899
e-mail: cpizarro@nemours.org*

Reference

1. Lange R, Vogt M, Hörer J, et al. Long-term results of repair of anomalous origin of the left coronary artery from the pulmonary artery. *Ann Thorac Surg* 2007;83:1463–71.