Anomalous origin of the left coronary artery from the pulmonary artery: late results with special attention to the mitral valve

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Abstract

Objective: Evaluate the late results of a uniform approach to the surgical management of children with anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA).

Methods: Between 1986 and 2007, 62 children with ALCAPA underwent surgery. The median age at operation was 16 months (range 10 days to 11 years). A uniform approach was applied, including (1) immediate surgery as soon as the diagnosis was established, (2) direct aortic reimplantation of the anomalous artery, when technically feasible (61/62, 98%), and (3) no concomitant mitral valve surgery, regardless of the severity of mitral regurgitation (59/62, 95%). The mean follow-up was 9.7 years (range 3 months to 21 years) and was 98% complete.

Results: There were six hospital deaths (9.7%). Left ventricular assistance was used in four patients; two died of related complications. The poor left ventricular ejection fraction was an incremental risk factor for early mortality (p = 0.043); severity of mitral regurgitation was not. There were two late deaths, yielding an actuarial survival rate of 86% at 15 years. Five patients underwent reoperation (mitral valve repair in three, coronary procedure in two); the actuarial freedom from reoperation was 89% at 15 years. Left ventricular function recovered in all survivors. In the 50 late survivors who did not undergo mitral surgery at initial operation, the severity of mitral regurgitation decreased in 58%, remained unchanged in 40% (of which 3 patients underwent reoperation for mitral valve repair) and worsened in 2%; at last follow-up, mitral regurgitation was absent or trivial in 42%, mild in 50%, moderate in 8% and severe in 0%.

Conclusions: (1) Early mortality is related to the severity of preoperative left ventricular dysfunction; it may be reduced by a careful use of postoperative cardiac support techniques. (2) Late results are satisfactory and left ventricular function always recovers. (3) Mitral regurgitation improves along with left ventricular function, but recovery may be incomplete and need reoperation. The data suggest that mitral valve surgery is probably not indicated at initial surgery, except in selected cases with a low potential of recovery (severe regurgitation with relatively well-preserved left ventricular function).

1. Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac malformation, which produces myocardial ischemia with left ventricular dysfunction and mitral regurgitation. The natural prognosis is very poor, but it has been changed by modern cardiac surgery. There were long controversies regarding the optimal technique of surgical repair and the ideal timing of operation [1]. A broad agreement has been reached to advocate (1) restoration of a two-vessel coronary system by direct aortic reimplantation of the anomalous artery [2–4] and (2) early surgery to promote rapid recovery of left ventricular function [5–7]. There remains, however, some discussion regarding the management of associated mitral regurgitation and the need for concomitant mitral valve repair [8–10].

During the last 20 years, a uniform protocol has been applied in our institution; this includes (1) early surgery as soon as the diagnosis is established, (2) direct aortic reimplantation of the anomalous artery, whenever technically feasible, and (3) avoidance of associated mitral surgery in the vast majority of patients. The present study was undertaken to evaluate the late results of this uniform approach, with special attention given to the fate of the mitral valve.
2. Materials and methods

2.1. Patient population

Permission to perform health records review was obtained from the Paris V university ethics committee. The need for individual consent was waived.

Between January 1986 and June 2007, 62 children (less than 18 years of age) underwent surgery for ALCAPA. One patient with associated hypoplastic left heart syndrome was excluded. There were 23 boys and 39 girls. Median age at operation was 16 months (range 10 days to 11 years). Forty-seven patients (76%) were less than 1 year of age. The median weight was 7.8 kg (range 2.7—31 kg).

Upon clinical presentation, 38 patients (61%) had persistent symptoms of congestive heart failure; 6 (10%) were critically ill (requiring ventilatory and inotropic support). The diagnosis, suspected on clinical and electrocardiographic data, was ascertained by echo Doppler evaluation alone in 42 patients (68%) and needed angiographic confirmation in the remaining cases.

The median preoperative left ventricular ejection fraction (as assessed by echocardiography) was 32% (range 10—75%). Thirty-six patients (58%) had a preoperative ejection fraction less than 30%. The mean preoperative left ventricular end-diastolic diameter indexed to normal for body surface area was 1.6 ± 0.25 (range 1.1—2.0).

Mitrail regurgitation was evaluated using echo Doppler examination. Preoperatively, the degree of mitral regurgitation was 0 to trivial in 4 patients (6.5%), mild in 28 (45%), moderate in 17 (27.5%) and severe in 13 cases (21%).

2.2. Surgical management

During the study period, a uniform surgical protocol was applied. This included three main principles.

2.2.1. Timing of surgery

Surgical repair was carried out as soon as the diagnosis was made. Surgery was thus undertaken on an urgent basis (within 48 h) in patients with persistent congestive heart failure or critical clinical status and on a semi- elective basis (within a few days) in the remaining children.

2.2.2. Surgical repair of the anomalous coronary artery

Direct aortic reimplantation of the ALCAPA was performed, whenever technically feasible.

Our standard technique has been recently described in detail [11]. The anomalous coronary artery was excised from its pulmonary origin, mobilized and reimplanted directly into the ascending aorta. This technique was used in 61 patients (98% of all patients). The anomalous coronary ostium was located in the right-handed facing pulmonary sinus in 44 cases (71%), the left-handed facing sinus in 14 patients (23%) and the proximal right pulmonary artery in 3 cases (4.5%).

In one patient (1.5%) in whom the artery was originating from the anterior non-facing pulmonary sinus, the standard technique could not be used and a long neo left coronary trunk was reconstructed to allow coronary revascularization (posterior pulmonary arterial flap and anterior autologous pericardial patch).

2.2.3. Management of the mitral regurgitation

In most patients (59/62, 95%), the mitral valve was not addressed at initial surgery, even in the presence of moderate or severe mitral regurgitation.

Mitrail valve repair was performed in only three patients with severe regurgitation (aged 15, 66 and 96 months) in whom there were, in addition to mitral annulus dilatation, evident ischemic lesions of the papillary muscles. One patient had shortening of elongated chordae; another had posterior annuloplasty using absorbable suture material; the last one had quadrangular resection of the posterior leaflet with posterior absorbable annuloplasty.

2.3. Follow-up and data analysis

Follow-up data were obtained during a 2-month closing interval (December 2007 and January 2008) by the physician or patient contact when the patients were not followed-up in our hospital. Information was obtained for all patients except one (98% complete).

Data were described as frequencies, medians with ranges and means with standard deviations. Crude ratios were given with 70% confidence intervals. Continuous variables were compared using independent t-test or Mann—Whitney U-test when data were not normally distributed. Pearson χ² or Fisher’s exact tests were used to determine differences when variables were expressed by dichotomous values. Estimates of time-related survival and freedom from reoperation were calculated using the Kaplan—Meier method. All analyses were performed using SPSS for Windows version 13.0 (SPSS Inc., Chicago, IL).

3. Results

3.1. Early results

3.1.1. Hospital mortality

Hospital mortality was defined as death within 30 days of operation or during the same hospital admission. There were six hospital deaths (9.7%, 70% confidence intervals 6—15%). The causes of death were: ventricular arrhythmias (2), neurologic damage (1), multi-organ failure (1) and untractable low cardiac output syndrome (2, both before the availability of left ventricular support). The median delay between operation and death was 9.5 days (range 1—22 days).

The following factors were predictive of hospital death: poor preoperative left ventricular ejection fraction (<30%) (p = 0.035), origin of ALCAPA from the left-handed facing pulmonary sinus (p = 0.026), operation before 1997 (p = 0.034) and use of left ventricular assist device (p = 0.043). The severity of preoperative mitral regurgitation was not identified as a potential risk factor. Among the 59 patients who did not undergo concomitant mitral surgery; there were 3 early deaths in the 32 children with no or mild mitral regurgitation (9.4—70% confidence intervals 4—18%) and 3 deaths in the 27 other patients with moderate or severe regurgitation (11.1—70% confidence intervals 5—21%).

3.1.2. Left ventricular assist device

A left heart assist device (with a centrifugal pump system) was used in four patients (6.5%). The median
duration of device implantation was 6.7 days (range 6—7 days).

Factors potentially associated with the need for assist device were: preoperative critical clinical condition ($p = 0.011$), poor left ventricular ejection fraction ($p = 0.05$), and long cardiopulmonary bypass time ($p = 0.029$).

All patients were successfully weaned from cardiac assistance, but two died from complications related to the device: cerebral hemorrhage in one and multi-organ failure in the other. The two other patients survived and had a normal left ventricular function at last follow-up.

3.1.3. Early morbidity
The median stay in the intensive care unit was 10.6 days (range 1—49 days). The median durations of ventilatory support and inotropic support were 7.2 days (range 0.5—41 days) and 7.6 days (range 1—48 days), respectively.

The sternum was left open in 16 patients (26%), for a median duration of 4.8 days (range 1—16 days).

3.2. Late results

3.2.1. Late survival and functional status
The mean duration of follow-up was 9.7 ± 6.6 years (range 3 months to 21 years). Among the 56 operative survivors, only one was lost to follow-up.

There were two late deaths (one sudden and one from cardiac failure during intercurrent lung infection) at 3 and 7 months postoperatively. The actuarial survival rate was 86% (95% confidence interval: 80—93%) at 15 years (Fig. 1).

At last follow-up, 45 patients (85%) were leading a normal, fully active life and 8 (15%) were in functional class II.

3.2.2. Late reoperations
The actuarial freedom from reoperation is shown in Fig. 2.

Five patients underwent reoperation (8.9%). Reoperations included mitral valvuloplasty (3), surgical angioplasty of the reimplanted left coronary artery (1) and mammary to left coronary bypass (1). Reoperations were performed after a median delay of 86 months (range 8 months to 18 years). No patient died at reoperation.

3.2.3. Late echocardiographic results
At last follow-up, the mean left ventricular ejection fraction was 63 ± 10% (range 35—75%) ($p < 0.00001$ compared to preoperative). The left ventricular end-diastolic diameter indexed to normal was 1.06 ± 0.10 ($p < 0.00001$ compared to preoperative).

3.2.4. Fate of the mitral valve
At last evaluation (53 patients), mitral regurgitation was absent or trivial in 21 cases (40%), mild in 28 (53%) and moderate in 4 patients (7%).

As previously stated, among the 13 patients who had preoperative severe mitral regurgitation, 3 underwent mitral valvuloplasty at the time of ALCAPA repair. The 3 patients survived and had mild residual mitral regurgitation at last follow-up.

The results regarding mitral valve function in the 50 survivors who did not undergo mitral surgery at initial operation are summarized in Fig. 3. Mitral regurgitation decreased in most cases (29/50, 58%), remained stable in 20 cases (40%) (of which 3 patients underwent reoperation) and worsened in only 1 patient (2%).

Three patients (with severe mitral regurgitation preoperatively) developed symptoms related to persistent severe mitral insufficiency after surgery and needed reoperation 8, 72 and 120 months after initial surgery. Chordal shortening in one patient and absorbable posterior annuloplasty in two was performed. At last follow-up, one patient had no regurgitation and two had mild residual insufficiency.

4. Discussion

4.1. Techniques of repair
Several surgical techniques have been advocated to repair ALCAPA, including left subclavian-left coronary artery anasto-
Fig. 3. Outcome of mitral regurgitation in the 50 survivors who did not undergo mitral valve surgery at initial operation. Three patients (*) underwent reoperation for mitral valve repair.

4.2. Mortality and risk factors for death

Recently published mortality rates after surgical repair of ALCAPA range from 0% to 16% [4—10]. It is shown in several reports (including the current study) that the main incremental risk factor for mortality is the preoperative severity of left ventricular dysfunction [2,7,13]. The severity of preoperative mitral regurgitation has also been proposed as an incremental risk factor [5], although this is not supported unanimously. In the present series, severe mitral regurgitation was not a risk factor.

Encouraging results have been achieved with cardiac mechanical support in the postoperative period, including extracorporeal membrane oxygenation or left ventricular assist device. These techniques must be used whenever weaning from cardiopulmonary bypass under stable hemodynamic condition is not possible. The full array of circulatory support should always be readily available, if required. In most cases, left ventricular function, even if extremely impaired preoperatively, recovers enough within a few days to allow successful weaning from cardiac assistance [4,8,14]. However, it should not be forgotten that pediatric cardiac assist devices carry their own mortality and morbidity. In the present study, two patients successfully weaned from cardiac assistance, died from related complications. We believe, as others [15], that careful intraoperative myocardial preservation, progressive weaning from cardiopulmonary bypass and appropriate postoperative inotropic support, should avoid the need for mechanical circulatory support in most cases. The indication for cardiac assistance should be carefully discussed on a case-by-case basis.

Late mortality is very low, particularly after recovery of the left ventricular function. However, during the recovery period, which may last several months, strict cardiac surveillance is necessary. Arrhythmias or cardiac failure may occur. Two of our patients, coming from developing countries, died a few months after surgery, before complete left ventricular recovery.

4.3. Fate of the mitral valve

The management of the regurgitant mitral valve at the time of ALCAPA repair remains controversial and variable. Most centers recommend not addressing mitral regurgitation, even severe, at initial surgery [1,6—8,10,15], whereas a few others advocate routine mitral valve repair [9,16]. Our own policy has been to avoid concomitant mitral valve repair, except in selected patients with evidence of irreversible damage to the papillary muscles. Our data following this policy show that (1) the early mortality rate is not influenced by the severity of preoperative mitral regurgitation, (2) the severity of mitral regurgitation decreases or remains stable in most patients, (3) reoperation for secondary mitral valve repair is seldom necessary.

On a theoretical basis, repairing the mitral valve where the myocardium is revascularized, seems logical; this should improve the early postoperative cardiac output, thus decreasing mortality and morbidity. However, this theoretical advantage faces major difficulties. Achieving effective mitral repair may be difficult in infants and young children; complex procedures on the papillary muscles and chordae are technically demanding; restrictive annuloplasty (the procedure established as optimal in adults with ischemic mitral regurgitation) cannot be used in infants because of the lack of growth potential. Performing complex mitral valve repair is time-consuming, thus increasing the ischemic time, which may be deleterious in patients with compromised myocardial function [8]. With all these considerations in mind, we think that a flexible attitude is appropriate.

In most patients with ALCAPA (particularly in infants), mitral regurgitation is related both to ischemic left ventricular dilatation with mitral annulus enlargement and...
to ischemic dysfunction of the papillary muscles. After successful coronary revascularization, left ventricular dimensions and function always improve and, even in most cases, recover to normal. This is associated with a reduction of mitral annulus size and, consequently, in the majority of patients, a decrease in the severity of mitral regurgitation. Our data support this hypothesis, particularly in patients with severe dilatation of the mitral annulus. The policy of not addressing mitral regurgitation at initial surgery, but rather waiting for ventricular remodeling to ensue is advocated by most centers [1,6–8,10]. When mitral regurgitation persists or worsens, the adequacy of coronary reimplantation must be ascertained [8]. Even after successful coronary revascularization and complete recovery of left ventricular function, mitral regurgitation may persist, due to irreversible lesions of the subvalvar mitral apparatus (elongation of chordae or fibrosis of papillary muscles). Reoperation for complex mitral repair or, sometimes, mitral valve replacement is then necessary, but can be performed with a low risk on an older child with normal left ventricular function.

There are, however, some patients with ALCAPA (usually children beyond infancy) in whom mitral regurgitation is severe despite a relatively well-preserved left ventricular function. The mechanism of mitral regurgitation is related more to ischemic lesions of the papillary muscles than to enlargement of the mitral annulus. Mitral valvuloplasty, in association with ALCAPA repair, is then indicated, because the likelihood of regression of mitral regurgitation with coronary revascularization alone is low [16]. Restrictive annuloplasty using absorbable material may be a satisfactory option.

5. Conclusion

The present study shows that direct aortic reimplantation can be performed in the vast majority of children with ALCAPA and provides satisfactory long-term results. Early mortality is related to the severity of left ventricular dysfunction and should be decreased by a wise use of postoperative cardiac assist devices. Left ventricular function always improves and usually recovers to normal. Our data support the policy not to address mitral regurgitation at initial operation (except in children with preserved left ventricular function), but reoperation for persistent mitral insufficiency may be necessary in a few patients.

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References


Appendix A. Conference discussion

Dr C. Mignosa

Dr Raisky

Dr C. Mignosa

But the thing that, I must say, needs to be probably better clarified is related to the mitral valve. As has been said, it is not necessary to treat the mitral valve electively. This is due to the fact that it is very complex, as we know, to repair such valves, mainly, when we are talking about infants. And on the other hand, we saw from the data that the improvement in the myocardial function gives recovery to the mitral valve.

But again, I would like to know if there are preoperative echocardiographic data that can drive our strategy and select patients who will require mitral valvuloplasty or not. I would like to have your comment on this point.
series, this was quite difficult to have this data, in the 1980s and it was one of the problem that we encountered to analyze these results.

The last key point is to appreciate the mitral regurgitation, I mean its mechanism, probably more important than its regurgitation volume. I think there are two groups. First group, could be called functional mitral regurgitation, due to ischemic lesion of the papillary muscle, and secondary dilated mitral annulus, both reversible after revascularization. The second group could be called organic mitral regurgitation, generally seen at follow-up with a preserved LV function. In this group, the function of the mitral valve is not improved by reimplanting the coronary because the lesions are due to anomalies of the subvalvular apparatus like elongation of the chordae, or huge fibrosis of the papillary muscle. These patients can probably benefit from a mitral repair, usually late after direct aortic reimplantation of the left coronary.