Long-term Results and Evolution of Mitral Regurgitation after Surgical Treatment for Anomalous Left Coronary Artery from Pulmonary Artery at Ho Chi Minh City Heart Institute

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ABSTRACT

Background:

Mitral regurgitation in anomalous left coronary artery from the pulmonary artery (ALCAPA) is secondary and caused by ischemia of papillary muscle. The study aimed to evaluate the long-term results of surgery and the evolution of mitral valve regurgitation after repair.

Methods

Retrospective review of all patients who underwent operation for ALCAPA with or without mitral repair at Ho Chi Minh City Heart Institute during the period 2000-2021.

Result

A total of 56 patients (male, 23; median age 1,67 y.o) were divided into two groups: infant ≤ 12 months (n= 25) and adults type(n=31 patients). Surgical reimplantation of the left coronary artery accounts for 43 cases for both groups. In group 2, Takeuchi's procedure was 3 cases and left coronary bypass in 6 cases. In both groups, 33 cases required mitral valve repair, 4 cases required replacement and 19 cases did not required intervention the mitral valve. There were 3 operative deaths (all in group 1) and no late deaths. The mean follow-up time after surgery was 92.5 ± 56.6 months. Only one patient lost to follow-up.The 52 survivors all had LVEFs above 55-60%. Overall cumulative survival at 5, 10, and 20 years was 94.6 ± 3%. 6/33 cases with initial valve repair had to have re-operation due to recurrent valve regurgitation (3 months-19 years after the initial surgery). At the time of final follow-up, excluding 3 cases of replace, the remaining 30 patients have mild to moderate mitral regurgitation. 19 patients without mitral intervention did not significantly change the degree of mitral regurgitation. Freedom from all causes of re-operation at 5 and 15 years was 94.4 ± 4% and 82.6 ± 8.6%. Freedom from re-operation for evolutive mitral regurgitation at 15 years was 84.8 ± 1.06% (group 1) and 81.8 ±1.16% (group 2), P= 0,045.

Conclusion

Surgical treatment for ALCAPA gives good long-term results. Evolutive mitral regurgitation after mitral surgery is more common in adults than in the infant group and requires continued periodic monitoring in follow-up.

Keywords: *ALCAPA, mitral regurgitation, reimplantation of the left coronary artery, redo*

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Background

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital heart defect, occurring in approximately 1 in 300,000 births (approximately 0.25-0.49% of congenital heart disease) and is a cause of myocardial infarction or sudden death in both children and adults. First described in 1933 by Edward Bland, Paul Dudley White and Joseph Garland, the Bland-White-Garland syndrome describes an ostium of the left coronary artery (LCA) originating from the pulmonary artery (PA). Physiologically, two months after birth, when the pressure in the PA suddenly drops, the left heart is not sufficiently nourished, causing necrosis and fibrosis of the endocardial myocardium. The consequence is reduced left ventricular function, and secondary mitral regurgitation. Clinical manifestations depend on whether severe left ventricular ischemia is compensated by the right coronary artery or not. Currently, based on clinical manifestation, ALCAPA is divided into two types: (1) infant type: children ≤ 12 months, this type has obvious clinical manifestations and is more severe than the adult type. In the infant type, left ventricular function is also severely impaired, severe mitral regurgitation due to anemia leads to nearly 90% early death in the first year if not operated promptly. (2) In the adult type, due to significant collateral circulation compensation from the right coronary artery, the consequences of myocardial anemia are also milder [1]. Definitive diagnosis is not difficult, but it is easy to miss if not thought of. Regarding surgical treatment, Neches et al [2] described the technique of directly re-implanting the left coronary artery root into the ascending aorta, also known as left coronary artery root node transfer, with good results, and this technique is widely accepted today. Some studies on short-term surgical results have shown that the main causes of early death are low cardiac output syndrome, ventricular arrhythmias, and severe residual mitral regurgitation. In the medium-term, left ventricular function has improved significantly but slowly and the rate of residual mitral regurgitation after surgery is still relatively high. In the long term, there are not many reports. We conducted this study to evaluate the long-term surgical results and progression of mitral regurgitation after surgical treatment for ALCAPA with mitral valve repair.

Patients and Methods

The study was conducted as a retrospective study of case series with longitudinal follow-up. We retrospectively reviewed 56 patients who underwent surgery to treat ALCAPA syndrome at the Ho Chi Minh City Heart Institute during the period from 2000 to 2021. Other coronary anomalies such as anomalous right coronary artery from pulmonary artery (ARCAPA) and coronary fistula were excluded from this study. Patients were divided into two groups: infantile (N1 = 25) and adult (N2 = 31). Pre-, intra- and post-operative data were collected using a data collection form. The two groups were compared using the Chi-square method and the mean of the two groups was compared using the T test. Cumulative survival rates and reoperation-free rates were calculated using the Kaplan-Meier method, and the two groups were compared using the Mantel-Cox log-rank. The study was approved and permitted to proceed by the Medical Ethics Council (Decision No. 594/QD-VT in 2023).

Results

Preoperative characteristics of the two groups are presented in Table 1. No patient in group 2 had a left ventricular ejection fraction (LVEF) below 30%, while 7/25 patients in group 1 had it. In contrast, the degree of left ventricular diastolic dilatation (LVEDD) was significantly greater in group 1. Moderate to severe secondary mitral regurgitation or more was more common in group 2 (77% vs. 64%).

Table 1: Preoperative characteristics of the two groups

Characterisrics	**Group 1 (n=25)**	**Group 2 (n= 31)**	***P@***
Mean Age (group 1= months; group 2= years) (min-max)	6.4 ± 2.8 (4-12)	22.2 ± 17.1 (1.33-60)	NA
Weight (kg) (min-max)	5.8 ± 1.14 (3.5-8.5)	38.4 ± 18,7 (8.5-68)	NA
Male	15	8	0.014
Cardiac US: LVEF	35.8 ± 12.3	59.1 ± 7.6	<0.001*
LVEF ≤ 30%	11	0	< 0.01
LVEF 31-50%	7	3	< 0.01
- LVEDD Z score	5.15 ± 1.4	3.9 ± 1.1	0.02
- MR: mild	2	2	0.625
moderate	7	5	
moderate to severe	4	11	
severe	12	13	
Direct signs: LCA ostium originated from PA + doppler color flow from LCA to PA	20 (80%)	18 (58%)	0.031
Indirect signs:			
- rich collateral circulation in the interventricular septum	16 (64%)	21 (67.7%)	0.784
- dilated right coronary artery	19 (76%)	23 (74.2%)	0.564
- fibrosis of the papilary muscle and endocardial associated with mitral regurgitation	20 (80%)	23 (74.2%)	0.754
Necrotic Q waves (DI, aVL, V3-V6) in ECG	14/18 (78%)	15/24 (62.5%)	NA
With P (ALCAPA) > 27	19 (76%)	NA	NA
MSCT confirms diagnosis	12 (48%)	28 (90.3%)	NA

LVEF: left ventricular ejection fraction; LVEDD: left ventricular end-diastolic diameter; MR: mitral regurgitation; PA: pulmonary artery. NA: non-applicable; @ Pearson Chi-square test ; * T-test

18/56 ECGs recorded before 2010 could not record results because the thermal paper was faded. MSCT is not routinely performed, and is mainly indicated in adults and cases where cardiac ultrasound images are still suspicious.

Table 2: Operative and post-op characteristics of the two groups

Characteristics	**Group 1 (n=25)**	**Group 2 (n=31)**	***P***
Technique: - LCA reimplatation	23	22	
- Takeuchi's tunnel	0	3	
- LCA bypass	0	6	
- Mitral valve repair	12	21	
- Mitral valve replace	0	4	
- No intervention on mitral valve	13	6	
Mean time of VC (hours)	149.4 ± 110.6	27.3 ± 29 (4-120)	< 0.001
Mean time stay in ICU (day)	10 ± 7.8 (1-33)	2.8 ± 2.4 (1-10)	< 0.001
Operative mortality (≤ 30 day)	03	0	0.047*

*Cochran's & Pearson Chi-square test

The majority of LCA roots originate from sinus 1, and 80% of patients had LCA reimplantation. 33 cases were repaired of mitral valve including posterior annuloplasty and anterior leaflet enlargement in group 1 and prosthetic ring annuloplasty (21 cases) or valve replacement (4 cases) in group 2. Surgical deaths in 3 cases were all in group 1. All 3 cases had pre-operative left ventricular function < 30%, all had LCA reimplantation, and 2 cases had mitral valve repair. Severe left ventricular failure and ventricular fibrillation were the main causes of death.

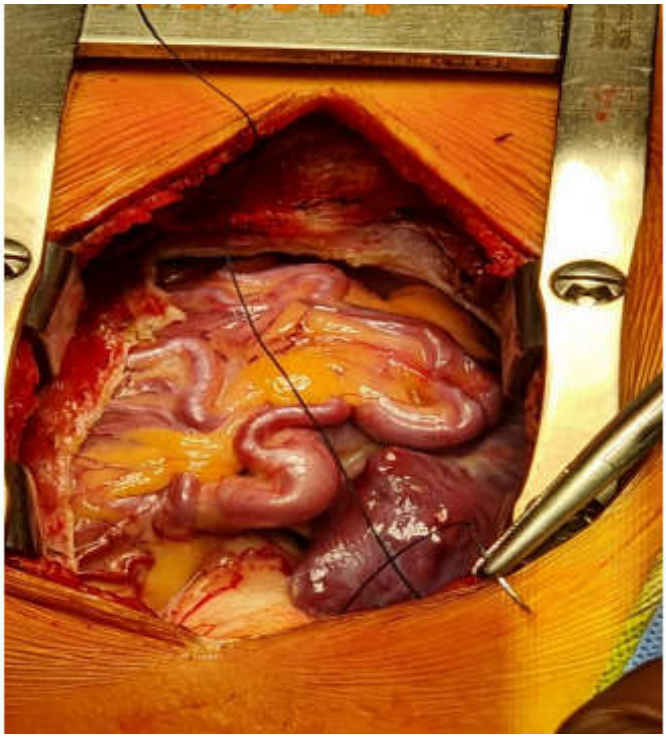

Figure 1: Right coronary artery dilatation in ALCAPA syndrome

Follow-up: the mean follow-up time was 80.45 ± 56.6 months (6-240 months) with a total follow-up of 4264 patient-years for all patients. Only 1 patient was lost to follow-up. At the final follow-up, no patient was in NYHA or Ross class III. After 2 years, the LVEDD Z-score value (45 patients) was mostly restored to normal: 1.08 ± 0.61. And the 52 surviving patients all had LVEF above 55-60%.

There were no late deaths. One patient with stenosis of the aortic ostium after Takeuchi surgery was detected on MSCT scan 7 years after surgery. Left ventricular function was preserved with LVEF # 55% and no obvious clinical symptoms. The patient underwent reoperation to reconstruct the tunnel using bovine pericardium. One adult case with fistulisation from the pulmonary artery tunnel (using autologous pericardium) was reoperated after 8 years, and was replaced with a Dacron patch. No case had pulmonary artery stenosis.

There were 6/33 cases with initial valve repair that had to have reoperation due to recurrent valve regurgitation. There were 2 patients in group 1 who were reoperated to re-repair at 3 months and 2 years after surgery. The other 4 cases in group 2 were reoperated for recurrent regurgitation: 3 cases had valve replacement (10, 15 and 19 years after the first surgery) and 1 case was re-repaired with ring (4 years after surgery).We excluded one case of progressive mitral regurgitation (no intervention on the valve at the initial surgery) requiring ring annuloplasty (7 years after surgery; this case was ALCAPA associated with tetralogy of Fallot). 19 patients without mitral intervention did not significantly change the degree of mitral regurgitation.Freedom from reoperation due to all causes after 5 and 15 years was 98 ± 1.9% and 94.3 ± 4.1%, respectively. Freedom of reoperation due to progressive mitral regurgitation after 15 years in group 1 was 84.8 ± 1.06% and in group 2 was 81.8 ± 1.16% (figure 2), a significant difference (P = 0.045; Mantel-Cox log-rank). 5, 10, 20 years survival rate for all patients was 94.6% (figure 3).

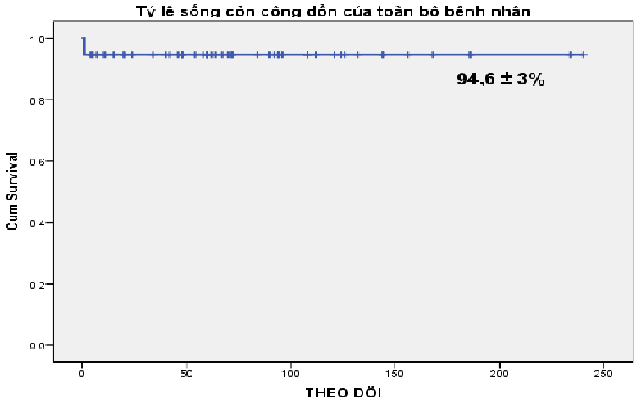

Figure 2: 5, 10, 20 years survival rate (KM) of all patients

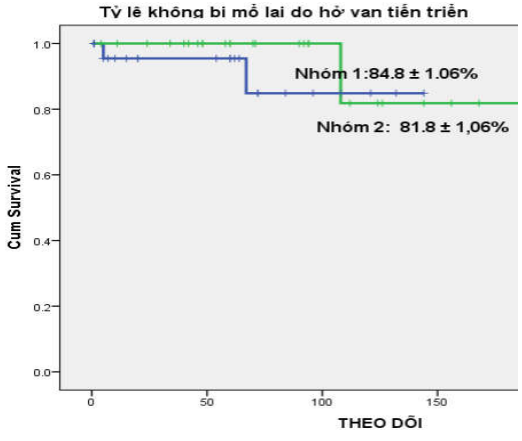

Figure 3: 15 years freedom of re-op due to progressive mitral regurgitation

Discussion

There have been many studies show that the earlier the surgical treatment, the better the prognosis thanks to the reestablishment of circulation in both coronary arteries. We prioritize the method of reimplanting the left coronary artery root (80%) for the majority of patients, including patients who undergo reoperation due to missed diagnosis. In cases where it is necessary to lengthen the left common trunk, we use the pulmonary artery wall as described by Turley [3] combined with the "trap door" technique when necessary. This lengthening technique (two-flap technique) allows reimplanting the left coronary artery root for most cases. And we do not use the pericardium for lengthening, nor do we use the Takeuchi technique for children like some other authors [4-7]. 3 cases of using Takeuchi's technique (one 8-year-old child and two adults were all performed before 2005, of which one case had fistulisation between tunnel and pulmonary artery (this patient was reoperated), and one case had a narrowed hole inserted into the aorta. This method has many complications, especially in children, such as pulmonary artery stenosis and tunnel fistula, so it is rarely used today. Furuta reported 3/11, and Hu 2/7 cases had severe complications after Takeuchi surgery. In this study, 6 patients aged older 50 underwent bypass surgery to the left anterior descending and sutured the root of ALCAPA. This technique is simpler and has less bleeding than re-implanting the left coronary artery root. After 2 years, almost all patients had normal left ventricular contractility and left ventricular diameter, demonstrating a spectacular recovery after re-establishing both coronary systems (dual coronary system). Thus, this surgical trend has been confirmed in terms of physiology and anatomy as well as bringing very good long-term results [3-7].

When is mitral valve intervention necessary? And how does mitral regurgitation progress after surgery? This issue is still controversial. Many authors only re-establish blood flow to the two coronary arteries and believe that mitral regurgitation will improve later and that only a few cases need to repair or replace the mitral valve later. On the contrary, many authors agree that intervention is needed when mitral regurgitation is severe due to ischemia or physical damage such as chordal elongation, valve leaflet cleft, etc. [8-14]. We also follow this point of view. And mitral valve repair techniques only include posterior annuloplasty and anterior leaflet enlargement in young children, and ring annuloplasty in older children and adults. Accompanying this is the repair of physical lesions such as valve leaflet cleft, chordal elongation, if any. We noted that the rate of severe mitral regurgitation in this study (25/56) was higher than in other foreign studies (Hu: 11/80; Yu: 18/111; Ismail: 7/29; Sasikumar: 3/46 patients). And the rate of needing reoperation for early or late mitral regurgitation was quite high at 6/33 cases; the majority of cases are in the adult group, similar to Naimo's study which was 3/11 [14]. With 6 patients needing reoperation for progressive mitral regurgitation, the average time to reoperation was 8.9 years (the earliest was 3 months and the latest was 19 years), showing that it is still necessary to continue regular follow-up after the initial surgery. At the final follow-up,

29/33 patients had only mild to moderate mitral regurgitation and no patients had severe regurgitation.The low surgical and late mortality, the spectacular recovery of left ventricular function, as well as the long-term results of progression of mitral regurgitation in this study showed the high effectiveness of surgical treatment to reconstruct both coronary artery systems. Furthermore, a meta-analysis of 907 patients from the European congenital heart surgery database concluded that mitral valve repair did not increase surgical mortality [9]. Only 4/19 patients with moderate to severe regurgitation in Kudumula's study had moderate regurgitation at the end of the study, although this author only repaired the valve for 4 cases [12]. In Yu's study, 27/34 patients with mitral repair had significant improvement in the degree of regurgitation, while only 19/37 patients without valve repair had improvement in the degree of regurgitation [14]. Thus, it can be seen that intervention for severe mitral regurgitation in ALCAPA is really necessary.

Conclusion

Surgical treatment for ALCAPA gives good long-term results. Postoperative progression of mitral regurgitation is more common in adults than in infants groups, so that requires regular follow-up after surgery. The rate of reoperation due to progression of mitral regurgitation is quite high but still acceptable. A larger sample size is also needed to accurately assess the long-term progression of mitral regurgitation.

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