Outcomes after Surgical Correction of Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

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SUMMARY

Background
The anomalous origin of the left coronary artery from the pulmonary artery is a strange congenital heart disease that produces myocardial ischemia with left ventricular dysfunction and mitral regurgitation of variable degree. The present techniques have modified the natural history of this disease through the re-establishment of a system of double coronary irrigation.

Objective
To evaluate the institutional experience in surgical correction of the anomalous origin of the left coronary artery from the pulmonary artery.

Material and Methods
Between February 2000 and May 2008, 13 patients with this diagnosis had an operation. Six of them were under 1 year old; mean: 2.5 years of age; range: from 45 days to 21 years of age. Before the surgery patients had: heart failure (61%), signs of myocardial infarction (69%) and moderate to severe cardiomegaly (84%). The average shortening fraction was 26.07% and there was moderate to severe mitral regurgitation in 46% of the cases. The diagnosis was confirmed in 11 patients by catheterization.

Takeuchi surgery was performed in one patient and in the remaining 12 the reimplantation of the left coronary artery in the aorta was carried out. Mitral valve surgery was performed in one patient. Six patients received perfusion and cardioplegia of the anomalous coronary artery through the pulmonary artery.

Results
At present, mortality is 0% with an average monitoring of 74-76 months. None of the patients had heart failure at the moment of leaving hospital. Cardiomegaly was reduced in 9 of 11 patients. The average post-operative shortening fraction was 39.92%. Mitral regurgitation decreased in all patients, excepts for one. None of the patients had severe mitral regurgitation in the remote post-operative period.

Conclusions
Results of corrective surgery of the anomalous origin of the left coronary artery from the pulmonary artery are excellent in all age brackets, with a significant improvement of myocardial function and a decrease of mitral regurgitation in most of the patients.

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MATERIAL AND METHODS
A retrospective and descriptive study, using the medical histories as an information source, was carried out. Data were collected and continuous categorical and numerical variables were defined. Tables and bar graphics were used for the description of data. Their statistical analysis was done with the program EpiInfo, version 3.3.2; a value of p < 0.05 was considered statistically significant for the comparison of means test.

Patients
13 patients were diagnosed as having ALCAPA between February 2000 and May 2008 at the Hospital de niños “Dr. Ricardo Gutiérrez” of the city of Buenos Aires. 8 patients were female and 5 male. All of them had an operation. According to age, at the moment of the surgical intervention, there were two groups: 6 patients under 1 year of age and 7 patients more than 1 year of age. The age range was from 45 days to 21 years and the mean age was 2.5 years.

Pre-operative evaluation
At the moment of pre-surgical evaluation, congestive heart failure in 8 of 13 patients (6 of them were under 1 year of age) was verified. The remaining 5 patients, with mild symptoms or without them, were the oldest of the entire group and all of them had more than 10 years of age. Eleven patients had moderate to severe cardiomegaly in the chest x-ray. The remaining 2, from 11 to 21 years of age, both asymptomatic, had cardiac silhouette within the normal boundaries.

In the electrocardiogram, pathological Q waves in the anterior side and in left precordial derivations were present in 9 ill patients.

The diagnosis was established through a color Doppler echocardiogram which identified the left coronary artery connected to the pulmonary artery. Likewise, the emergency place of the anomalous coronary artery, presence of intercoronary collaterals, associated intracardiac or extracardiac pathology, shortening fraction (SF) and degree of mitral regurgitation (MR) were evaluated.

The mean shortening fraction in patients under 1 year of age was 15.83% with a standard deviation of 2.40%. In patients more than 1 year of age was 34.85% with a standard deviation of 2.40%. In 9 patients, 6 patients under 1 year of age and 3 patients more than 1 year of age. The age range was from 45 days to 21 years and the mean age was 2.5 years.

Table 1. Pre-surgical characteristics of patients

<table>
<thead>
<tr>
<th>P</th>
<th>Sex (Gender)</th>
<th>Age (m)</th>
<th>HF</th>
<th>Q waves</th>
<th>Cardiomegaly</th>
<th>SF %</th>
<th>MR</th>
<th>Collat.</th>
<th>Origin of the LCA in the PA</th>
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<tr>
<td>1</td>
<td>F</td>
<td>144</td>
<td>No</td>
<td>Yes</td>
<td>Moderate</td>
<td>32</td>
<td>Mild</td>
<td>Yes</td>
<td>Posterior</td>
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<td>2</td>
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<td>Severe</td>
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<td>Mild</td>
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<td>3</td>
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<td>Mild</td>
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<td>Left posterior</td>
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<tr>
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<td>7</td>
<td>Yes</td>
<td>No</td>
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<td>Yes</td>
<td>Left lateral</td>
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<td>Yes</td>
<td>Yes</td>
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<td>Moderate</td>
<td>No</td>
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<tr>
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<td>M</td>
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<td>Yes</td>
<td>No</td>
<td>44</td>
<td>No</td>
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<tr>
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<td>30</td>
<td>Yes</td>
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<tr>
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<td>No</td>
<td>No</td>
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<tr>
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<td>M</td>
<td>31</td>
<td>Yes</td>
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<tr>
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<td>M</td>
<td>3</td>
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<td>Yes</td>
<td>Severe</td>
<td>15</td>
<td>Severe</td>
<td>No</td>
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A 30-month patient had severe MR with dysplastic and thickened mitral valve and with two fissures in the anterior leaflet. Annuloplasty and fissure closure before left coronary reconnection was performed.

RESULTS
Mortality in the short and long terms was 0%. Mechanical circulatory support was not required in none of the cases. In 2 patients, both under 1 year of age, the thorax was closed due to hemodynamic instability and severe cardiomegaly after the output of extracorporeal circulation.

The monitoring was carried out through scheduled interviews at the hospital office after 15 days, 1 month, 3 months and then every 6 months after leaving hospital, during an average time of 74-76 months. Also, in the intervals among consultations, each family got in touch with its respective pediatric cardiologist.

During the whole period of post-surgical monitoring, the general clinical condition with special emphasis in the cardiovascular aspect through a questioning and a detailed semiology was evaluated.

All symptomatic patients improved significantly after surgery. None of them had heart failure during the remote monitoring.

From 11 patients that had preoperative cardiomegaly in the chest x-ray, in 9 it was reduced. In the remaining 2, there was no important change in the cardiothoracic index, although both had clinically improved and their left ventricular function was normal.

In each scheduled consultation, a transthoracic echocardiography was performed. The post-operative SF 6 months after leaving hospital was 39.92% on average, with a standard deviation of 5.67%. In all the cases, the postoperative SF was greater than the pre-surgical one and it was always more than 30%. The mean was 36.50% in patients under 1 year of age, with a standard deviation of 3.72% in comparison with a pre-surgical mean of 15.83% (p < 0.0001). In patients more than 1 year of age, the mean was 42.85% with a standard deviation of 5.58% against a pre-surgical value of 34.85% (p = 0.0032). Theses data are summarized in Figure 1.

In patients with MR, this decreased in all of them, excepts in one. None of them had severe MR 6 months after leaving hospital; in two cases it was moderate and in the remaining 11 it was mild or it did not exist. Valvuloplasty performed in a 30-month boy reduced the regurgitation degree from severe to mild. There was a patient in which MR got worse from mild, before surgery, to moderate in the post-operative period. The left coronary artery was permeable in its entire path. These data are summarized in Figure 2.

Hemodynamics, effort test and myocardial perfusion studies were used as complementary methods. Only one patient required cardiac catheterization to confirm the permeability of the left coronary artery, since in the remaining 12 cases it was verified by echocardiography. Five post-operative effort tests were normal and two of four perfusion studies were also normal, whereas the remaining two showed a significant improvement of myocardial perfusion.

There were no reoperations during monitoring.

DISCUSSION
In most of the children still on milk, this pathology is presented as congestive heart failure with high mortality without treatment. The lack of intercoronary collateral flow causes severe ventricular ischemia. When the ductus is closed and the pulmonary resistances fall, the blood theft from the pulmonary artery favors ischemia. (3) Older children and adults with this heart disease are asymptomatic, since they have a plenty network of intercoronary collaterals and, sometimes, an ostial stenosis of the left coronary artery which limits the blood theft and increases the myocardial perfusion pressure. (2, 4)

A continuous spectrum, which goes from the myocardial infarction and the congestive heart failure in children to the heart murmur in MR in an asymptomatic patient with late presentation that may be complicated with the development of syncope and sudden death, is made up.

The electrocardiogram shows signs of ischemia or left anterolateral infarction, with profound Q waves,
inverted T waves and ST segment depression in I, aVL and left precordial derivations. The differential diagnosis among ALCAPA, myocarditis and dilated cardiomyopathy may be difficult. A formula is used to calculate the probability of existence of ALCAPA, in which it is equal to $12d + 24s - w$, where “d” shows the depth in millimeters of Q wave in aVL, “s” ST segment amplitude in millimeters in aVL and “w” the width in millimeters of Q wave in I. When this value is more than 27, ALCAPA diagnosis with a 100% of sensitivity and a 96% of specificity is established. (5, 6)

The two-dimensional and color Doppler echocardiograms may show the origin of the anomalous coronary artery, apart from the dilated right coronary artery and an entrance flow in the pulmonary artery. This flow may appear in two different patterns: continuous with systolic reinforcement in older children with several dilated intercoronary collaterals or late systolic in patients with scarce collaterals. (6)

There is global hypotomility with variable degree in the left ventricle, with increased end-diastolic volume. Color Doppler allows us to show the degree of MR.

This method has limitations in special occasions. High pulmonary pressure, proper in newborn babies or in children with left ventricular failure, generates an antegrade blood flow in the anomalous coronary artery, with scarce dilation of the right coronary artery. In adult patients, the demonstration of the anomalous origin may be more difficult, therefore the detection of dilated vessels in the anterior interventricular septum should trigger suspicion and guide the echocardiographic diagnosis step by step. (7)

Cardiac catheterization is the most effective method to establish the diagnosis with certainty. The aortography shows the emergency of an only coronary artery, the right one. Its selective injection allows us to observe its dilation and it presents a sinuous path. Intercoronary collaterals may be observed. Their contrast darkens the pulmonary artery. There exists an oximetry protuberance in the pulmonary artery due to the short circuit, with a relationship of pulmonary to variable systemic flow and generally with little significance. In cases with a minimum collateral circulation, this result may not be presented.

High pulmonary pressure in newborn patients may show antegrade flow in the left coronary artery when injecting contrast in the pulmonary artery. The left ventriculography shows dilation, hypokinesia and, frequently, MR with variable degree.

Nowadays, surgical indication is clear: the patient with confirmed ALCAPA diagnosis should undergo surgery. Early age should not be a risk factor to suspend surgery. Stable patients, regardless of their age, should undergo surgery immediately. The postponement of the surgical intervention implies the risk of worsening the clinical condition, in children still on milk and in older asymptomatic patients in which this pathology may derive in sudden death preceded or not by angina pectoris symptoms, heart failure or arrhythmias.

Although during decades, several surgical techniques for the treatment of this pathology were described, nowadays there is a general consensus regarding the superiority of the re-establishment of a system of double coronary irrigation over the ligature of the anomalous coronary artery. (8) These techniques produce a positive impact over the natural history of this disease, with low mortality and significant improvement of the left ventricular function. The possible mechanisms that underlie this improvement are hyperplasia and myocyte hypertrophy, apart from the recovery of the hibernating myocardium by chronic hypoperfusion.

This reorganization is slow and a progressive reduction of the left ventricular end-diastolic and end-systolic diameters is observed, even 6 months after surgery. (9) Although the ligature of the left coronary artery improves the clinical manifestation in emergency cases, (10) it presents an important decrease of the survival index in the long term. (8)

There is a wide variety of techniques to re-establish the system of double coronary irrigation. Direct reimplantation into the aorta is the chosen technique in most of the cases due to the increasing experience in the use of the coronary arteries in the transposition of the great vessels. Extracorporeal circulation with antegrade perfusion of the left coronary artery through the cannulation of the pulmonary artery is used. When both coronary territories are perfused, body temperature falls up to reach moderate or profound hypothermia, according to the chosen strategy. The aorta is clamped and antegrade cardioplegia in the aortic and pulmonary roots is administered. This guarantees a uniform myocardial protection in both ventricles (11, 12). The left coronary artery from the pulmonary artery with a button of surrounding tissue is disconnected and it is reimplanted in the aortic root.

In some cases, different types of tunneling of the left coronary ostium to the aorta may be useful. Tashiro et al. (13) described the preparation of a tunnel using a ring of tissue from the pulmonary artery from which the coronary artery emerges. Then, it is anastomosed to the posterior side of the aortic root. When the
anomalous coronary artery emerges, it goes down in the left posterolateral side of the pulmonary artery. The rectangular fragment of the wall of such artery, which goes up to the beginning of the left branch, may be used. Such fragment is used to produce a tunnel of autologous tissue which is connected to the aorta. (14)

In infrequent cases in which the coronary artery emerges from the anterior side of the pulmonary artery, tunneling using a button of pericoronary tissue with a portion of aortic tissue partially removed from the root may be useful. (15) Takeuchi et al. described the transpulmonary reconnection of the left coronary artery in the aorta in 1979 which consists in the creation of an aortopulmonary window and its connection to the coronary ostium through an intrapulmonary tunnel. (16)

These techniques have risk of obstruction in the tunneling. Takeuchi’s surgery may cause variable degrees of obstruction at the output of the right ventricle and also residual connections between the pulmonary artery and the lumen of the coronary artery.

The anastomosis of the left coronary artery to other arterial vessels or through venous bridges may be performed with or with no extracorporeal circulation and through multiple pathways of thoracic approach. However, it is technically more difficult in children still on milk and the obstruction due to torsion is still an important disadvantage. (17) MR has as a substrate, the dilation of the left ventricle with the consistent dilation of the mitral ring, ischemia or papillary muscle infarction and the dyskinesia of the left ventricular free wall. Although some groups carry out mitral plastic surgery in all patients with moderate to severe mitral regurgitation, (18) nowadays there exists a consensus in which valvuloplasty should not be practiced routinely. (19) In most of the cases, just with the correction of ALCAPA, MR is reduced in an important degree, with progressive improvement of the valvular continence even 1 year after the surgery. (9) In case mitral regurgitation remains, catheterization to discard the obstruction of the reconnected coronary artery is advisable to be done. (20) In those particular cases in which there is irreversible ischemic damage of the papillary muscles or some fissures in the leaflet, mitral plastic surgery should be considered. In the remaining patients, the degree of regurgitation decreased evidently with no need of any procedure on the valve.

In our series, cardiovascular mechanical support was not necessary, although it constitutes a valid method in patients with severe worsening of the myocardial function.

CONCLUSIONS

Results of corrective surgery of the anomalous origin of the left coronary artery from the pulmonary artery are excellent in all age brackets, with a significant improvement of the myocardial function and a decrease of mitral regurgitation in most of the patients.
1. Brooks HS. Two cases of an abnormal coronary artery of the heart arising from the pulmonary artery: with some remarks upon the effect of this anomaly in producing cirsoid dilatation of the vessels. J Anat Physiol 1885;20:26-9.


