

## Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery: Diagnosis and Postoperative Follow Up

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### Abstract

**Background:** Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital malformation that presents a diagnostic challenge to the pediatrician and pediatric cardiologist. Although surgical repair is always indicated, the optimal technique has yet to be determined.

**Objectives:** To review our experience with the diagnosis of children with ALCAPA and to assess short to midterm surgical results.

**Methods:** Between 1992 and 1998, 13 infants and children (2 months to 15 years) were treated for ALCAPA at our medical center. Eight were diagnosed during the first year of life; all were symptomatic and had severe dysfunction of the left ventricle. The five patients diagnosed at an older age had normal myocardial function. Diagnosis was established by echocardiography alone in seven patients; six required catheterization (one infant and all older patients). Surgery was performed in 12 patients to establish dual coronary artery system: 7 underwent the Takeuchi procedure and 5 had re-implantation of the anomalous left coronary artery.

**Results:** One infant died shortly after diagnosis before surgical repair was attempted, and one died postoperatively. Four patients required additional surgery: three for late complications of the Takeuchi procedure and one valve replacement for mitral insufficiency. Recent evaluation revealed good global left ventricle function in all patients except for one, who is still within the recovery phase and shows gradual improvement. However, most patients who presented with severe myocardial dysfunction upon diagnosis still display abnormal features such as echo-dense papillary muscles or evidence of small akinetic

segments. In this group, early repair was associated with faster myocardial recovery.

**Conclusions:** The diagnosis of ALCAPA remains a clinical challenge to the pediatrician and cardiologist. Diagnosis can be established echocardiographically, and early diagnosis and treatment may lead to faster myocardial recovery. The preferred surgical method appears to be re-implantation of the ALCA. The chance for good recovery of global ventricular function is high even in the sickest patients, nonetheless abnormal myocardial features can be identified even years after surgery.

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Anomalous origin of the left coronary artery from the pulmonary artery is a rare congenital heart lesion that, if untreated, results in high mortality, especially in symptomatic patients [1-3]. The associated clinical entity is characterized by chronically ischemic, frequently hypocontractile and potentially revivable, myocardium. Before birth, the pressure in the aorta and the pulmonary artery is equal and the oxygen saturation in the pulmonary and systemic circulation is similar. Therefore, while *in utero*, the anomalous coronary is supplied by antegrade flow of relatively saturated blood. Immediately after birth and during the first weeks of life, symptoms are rare, reflecting the elevated pulmonary vascular resistance that persists during the neonatal period. However, when pulmonary vascular resistance falls, most infants become symptomatic; this process takes place several weeks or months after birth [4]. Asymptomatic older patients carry the risk of sudden death, and in one series 80% died suddenly at a mean age of 35 [1,5,6].

The treatment of choice is to establish a dual coronary artery system, which can be achieved by the use of extracardiac arterial blood supply, re-implantation of the

ALCAPA = anomalous origin of the left coronary artery from the pulmonary artery

ALCA = anomalous left coronary artery

anomalous coronary ostia, or by creation of an aorticopulmonary window with an intrapulmonary baffle (Takeuchi procedure) [7]

This study reviews our clinical experience with 13 consecutive patients who were diagnosed and treated for ALCAPA at the Schneider Children's Medical Center of Israel.

## Patients and Methods

### Patients

Thirteen patients with ALCAPA were treated at our hospital between July 1992 and December 1998; their ages ranged from 2 months to 15 years [Table I]. Eight were diagnosed during the first year of life. All infants were symptomatic, presenting with respiratory symptoms or failure to thrive. One infant with a history of failure to thrive presented with malignant arrhythmia. The older patients were referred to a cardiologist for the evaluation of a cardiac murmur heard on routine examination.

### Diagnosis

All patients underwent echocardiographic evaluation. Diagnosis by echocardiography was accepted only if color Doppler indicated reverse blood flow within the left coronary artery and into the pulmonary artery. Using these criteria, cardiac catheterization was performed in six patients to further establish the diagnosis.

### Surgery

Twelve patients were operated on. One patient died shortly after diagnosis after developing multi-organ failure. The surgical technique was chosen to best fit the anatomy and was performed shortly after diagnosis. When performing the Takeuchi procedure, the intrapulmonary

artery baffle was constructed with a flap of the patient's own pulmonary artery. The pulmonary artery was then roofed with autologous pericardium.

### Follow-up

Postoperative follow-up was conducted at regular intervals (1–3 months initially and yearly later on), when physical examination and echocardiographic evaluation were routinely performed.

## Results

### Clinical evaluation

All patients diagnosed during the first year of life were symptomatic. All underwent an initial echocardiographic evaluation: ALCAPA was identified immediately in five infants. In two infants initially diagnosed with myocarditis, ALCAPA was identified at a later echocardiographic examination. All infants had severe dysfunction of the left ventricle upon diagnosis. One died shortly after diagnosis, before surgical repair was attempted, from severe heart failure and arrhythmia leading to multi-organ failure and severe neurological damage. Older patients had preserved myocardial function, and two of them were followed and treated for suspected cardiomyopathy. Catheterization was performed in all older patients, as well as in one infant to verify the echocardiographic evaluation after an initial diagnosis of myocarditis.

### Surgical repair

Surgery was performed shortly after diagnosis, and the surgical technique to best fit the anatomy was chosen. The Takeuchi procedure was performed in seven patients and re-implantation of the anomalous coronary into the aorta in five. One infant died of uncontrolled sepsis and respiratory failure one month after surgery. Four patients required additional surgery. Valve replacement for insufficiency was performed in one patient with structurally abnormal mitral. One patient who underwent the Takeuchi procedure developed occlusion of the tunnel requiring its reopening; the same patient developed progressive narrowing of the main pulmonary artery that necessitated surgical augmentation of the stenotic area. A second patient with the Takeuchi procedure developed increasing tunnel leak into the PA, which was closed surgically. A third patient who underwent the Takeuchi procedure at another hospital was identified as having occluded tunnel, and revascularization of the left main coronary was performed using the left internal mammary artery.

### Follow-up

The most recent evaluation revealed good global left ventricular function in all patients operated on prior to 1998, with normal fractional shortening as assessed by M-mode. None of these patients is symptomatic or receives medications (NYHA class I). One patient is still within the recovery phase and is showing gradual improvement. However,

PA = pulmonary artery

Table I. Results of surgical treatment in 13 patients with ALCAPA

Age	Year of surgery	Presenting symptoms	Diagnostic methods	Before operation	At last follow-up	Type of surgery
10 yr	1992	Murmur	CATH	Good	Good	Re-implantation
9 yr	1992	Murmur	CATH	Good	Good	Takeuchi
5 mo	1993	Respiratory	ECHO	Bad	Good	Takeuchi
14 mo	1994	Murmur	CATH	Good	Good	Re-implantation
6 mo	1994	Respiratory (FTT)	CATH	Bad	Good	Takeuchi
3 mo	1996	Respiratory	ECHO	Bad	Good	Re-implantation
4 mo	1996	Respiratory	ECHO	Bad	Good	Takeuchi
15 yr	1997	Murmur	CATH	Good	Good	Takeuchi
2 mo	1997	Respiratory	ECHO	Bad	Good	Takeuchi
11 mo	1998	Cardiac arrest, arrhythmia*, FTT	ECHO	Bad	Good	Takeuchi
4 mo	1998	Respiratory	ECHO	Bad	Good	Re-implantation
7 mo	1998	Myocarditis	ECHO	Bad	Died	Re-implantation
13 yr	1998	Respiratory	CATH	Good	Good	Takeuchi

\* Malignant arrhythmia.

CATH = definitive diagnosis made by cardiac catheterization, ECHO = definitive diagnosis made by echocardiography, FTT = failure to thrive, murmur = murmur heard on routine examination, re-implantation = direct implantation of the left coronary ostia, respiratory = dyspnea as presenting symptom, Takeuchi = aortopulmonary artery tunnel procedure.

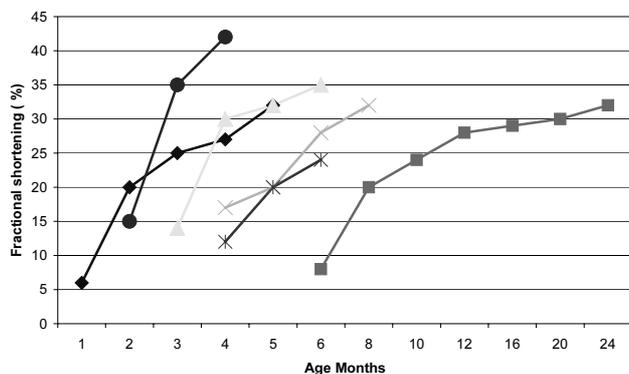


Figure 1. Cardiac function recovery after surgery in the infant group, evaluated by echocardiography and expressed in fractional shortening (FS %). The Y-axis presents FS in %; the X-axis presents age in months and period of follow-up until normal function is reached. The first point presents the age of presentation and cardiac function before surgery in each patient.

most patients who presented with severe myocardial dysfunction upon diagnosis still display abnormal features such as echo-dense papillary muscles or evidence of small akinetic segments. Early repair was associated with faster myocardial recovery [Figure 1].

## Discussion

ALCAPA is a rare congenital heart defect that results in high mortality if not treated surgically. Although the associated clinical picture is well characterized and includes congestive heart failure, cardiomegaly and abnormal ECG pattern [1–3], early diagnosis remains a clinical challenge to the pediatrician because the initial symptoms in infants are nonspecific while most older children do not display any symptoms at all. The pediatric cardiologist also faces a diagnostic challenge since the ability to demonstrate coronary anatomy and flow in ultrasound is mandatory for positive identification of this condition. Our clinical study clearly shows that careful echocardiography can provide accurate and sufficient diagnosis especially in infants, while previous studies indicated unreliable visualization of the origin of coronary arteries [8–11].

Our study suggests that in our young patients, all of whom presented with decreased cardiac function, early diagnosis and surgery may lead to faster recovery. The pathophysiology of the clinical manifestation in ALCAPA was proposed by Edwards [12]. In the neonate, blood will flow from the pulmonary artery into the anomalous left coronary artery as a result of the high pulmonary vascular resistance. As pulmonary resistance and pressure falls, less blood flows from the pulmonary artery into the left coronary. Blood supply will then depend on intercoronary collaterals from the right coronary. It can be speculated that infants who develop early signs of congestive heart failure have insufficient intercoronary collaterals and therefore do not have sufficient blood supply for myocardial function [13]. It is therefore logical to assume that in such patients early diagnosis will allow a shorter period of

myocardial ischemia and that early establishment of normal left coronary flow will result in a faster and more complete recovery. Such a process is suggested by our data, although a larger group of patients will be needed for statistical significance. In other cases these collaterals may be sufficient or may develop over a short period, allowing adequate myocardial blood supply. Such patients may remain asymptomatic for years, maintain good myocardial function, and can produce a significant left to right shunt [14].

The goal of modern surgical management of anomalous left coronary artery is to establish a dual coronary system with long-term patency. This can be achieved by the use of native tissue and by maintaining the ability for growth of the coronary ostia and arteries. Several methods have been described for establishing left coronary continuity with the aorta, such as creation of an aortocoronary bypass and ligation of the left coronary origin, direct implantation of the left coronary artery to the aorta, and creation of an aortopulmonary tunnel.

Several techniques can be used for the creation of an aortocoronary bypass. Saphenous vein graft, subclavian artery and left internal mammary have all been used as conduits between the aorta and the anomalous coronary. The use of saphenous vein has been limited in babies by the small diameter of this vessel and by its tendency for late occlusion [15]. The subclavian artery has been used to revascularize the left coronary, although several technical difficulties including significant size mismatch of both arteries, kinking of the subclavian artery, and late anastomosis stenosis have been described [16]. The use of the left internal mammary to provide oxygenated blood to the left coronary artery can be used in older children, whereas it is probably too small to be used in infants and its growth ability has not yet been established.

Direct implantation of the left coronary ostia was first performed by Neches et al. [17]; the first series was described in older patients by Grace et al. [18] and did not include infants. Later series used this technique in young babies with increasingly good results [18–20].

In 1978, Takeuchi and colleagues described the creation of a tunnel within the pulmonary artery as well as an aortopulmonary window to direct oxygenated aortic blood into the ostium of the anomalous coronary. The tunnel was formed from a flap of the pulmonary artery. A major theoretical advantage of this procedure is the use of *in situ* tissue, which maintains its growth potential. The defect in the pulmonary artery was then repaired using a pericardial patch.

The overall survival rate after dual coronary repair in our series was 92%, a rate comparable to reports from leading tertiary care centers in the world [21]. According to our experience, the preferred method appears to be direct implantation of the anomalous coronary to the aorta. Indeed, the only infant who did not survive after surgery underwent this type of operation, but this death occurred

more than a month after surgery and was caused by infection resulting in severe lung disease. This patient, the oldest among the infants undergoing surgery, was severely underweight because of heart failure, and received steroids for the presumed diagnosis of myocarditis prior to surgery. Forward flow into his implanted left coronary was demonstrated in several echocardiographic studies indicating a satisfactory technical result. The other patients with implanted coronary had no complications on follow-up. On the other hand, three of the seven patients with the Takeuchi procedure developed several complications including supravulvar pulmonic stenosis, baffle leak into the pulmonary artery, and tunnel occlusion, requiring additional surgical interventions. Similar problems have been described in several other series [19,21,22]. Therefore, our experience indicates that when anatomically feasible, the direct implantation appears to be superior to any other technique.

Several conclusions can be drawn from our experience. Firstly, in spite of the great advancement in modern medical methodology, the diagnosis of a child with ALCAPA continues to be a diagnostic challenge. Secondly, echocardiography can provide an accurate and definite diagnosis. Thirdly, implantation of the anomalous coronary provides better long-term results with fewer complications. Fourthly, the high survival rate and good postoperative myocardial recovery even in patients with markedly depressed myocardial function do not justify consideration of cardiac transplantation. Lastly, early surgery in infants with ALCAPA may be associated with a faster recovery rate.

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*The voyage of discovery is not in seeking new landscapes but in having new eyes.*

*Marcel Proust*