

# Anomalous Origin of the Left Coronary Artery from the Right Pulmonary Artery with Intramural Aortic Trajectory. Clinicosurgical Diagnostic Implications

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**Objective** - Anomalous origin of the left coronary artery from the right pulmonary artery (AOLCARPA), is a rare entity that is usually associated with other defects. Of the 20 cases of AOLCARPA reported in the literature, 14 (70%) had associations. We describe four patients with AOLCARPA without associated defects, but with a peculiar intramural aortic trajectory.

**Methods** - Fifty-five patients with anomalous origin of the left coronary artery were operated upon at INCOR-FMUSP. Four of the patients had the anomalous origin from the right pulmonary artery (RPA) without associated defects but with intramural aortic trajectory. Clinical and laboratory examinations were analyzed, as well as surgical findings.

**Results** - All patients had congestive heart failure (CHF) and 3 also had angina pectoris. Two patients had a murmur of mitral regurgitation, signs of myocardial infarction on the ECG and cardiomegaly. The shortening fraction varied from 9% to 23%. The hemodynamic study confirmed the diagnosis of anomalous origin of the coronary artery, but the intramural trajectory and the origin from the RPA were established only at surgery. In 3 patients, the technique of side-to-side anastomosis was performed with a good outcome. One patient, who underwent end-to-side anastomosis, died 6 months after the surgery.

**Conclusion** - Association with other defects usually occurs in the AOLCARPA, and the intramural aortic trajectory is difficult to clinically diagnose but easy to surgically correct.

**Key words**- anomalous origin of the left coronary artery, right pulmonary artery

Anomalous origin of the left coronary artery from the pulmonary arterial tree (AOLCAPA) is a rare anomaly with a high mortality rate, even in the first months of life. Survival clearly relates to the magnitude of the collateral arteries originating from the right coronary artery to the left coronary artery<sup>1,2</sup>.

AOLCAPA is rarely associated with other congenital defects, unless the right pulmonary artery (RPA) takes part in the anomaly. The presence of associated defects in 70% of the 20 cases of origin of the left coronary artery from the RPA reported in the literature is well known<sup>3-21</sup> (table I).

In some of these cases<sup>12,15,17</sup>, in addition to the anomalous origin from the RPA, a rare anatomical characteristic – the intramural aortic course – complicated the clinical diagnosis but, on the other hand, facilitated the surgical correction.

The objective of this study is to report our experience with this anatomical peculiarity, rarely described in the literature<sup>12,15,17</sup>.

## Methods

Between 1987 and 1997, at the Instituto do Coração of the Medical School of the University of São Paulo, 55 patients with AOLCAPA underwent the surgical correction for the isolated defect. Four (7.2%) patients had the anomalous origin directly from the RPA with intramural aortic trajectory, comprising the objects of this study.

Three patients were males and one was female, and ages ranged from 3 to 15 months (mean = 6.75 months). All infants showed clinical evidence of congestive heart failure (CHF) at the initial examination. Three of the patients showed great irritability as an expression of the clinical syndrome of angina pectoris.

All clinical findings and laboratory tests were reevaluated, including the electrocardiogram (ECG), the chest X-ray and the echocardiogram. The surgical findings, as well as the evolutionary characteristics, were revised. The opera-

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Table I - Reports in the literature of anomalous origin of the left coronary artery and of the circumflex* coronary artery from the right pulmonary artery					
Cases	Author	Year	Age	Associated defects	Death
1	Masel LF	1960	3 w	TF	Yes
2	Rao BNS	1970	3 y	VSD	Yes
3	* Honey M	1975	13 y	CoAo	-
4	Doty DB	1976	10 m	-	-
5	* Ott DA	1978	8 y	-	-
6	Driscoll DJ	1982	3 w	CoAo+ AoS +AVC+PDA	?
7	* Daskalopoulos	1983	13 m	CAT	Yes
8	Bharati S	1984	3m	CoAo	Yes
9	Hamilton JRL	1986	3m	-	-
10	Atik E	1988	5m	-	-
11	Levin SE	1990	7 sem	CoAo	Yes
12	Levin SE	1990	7m	CoAo	-
13	Henglein D	1990	8m	VSD	Yes
14	Atik E	1991	15 m	-	-
15	Igarashi H	1993	12 m	MR	-
16	Turley K	1995	6m	-	-
17	Tanaka SA	1996	12 y	MR	-
18	Sarris GE	1997	9m	LHH	-
19	* Sarioglu T	1997	10 y	CoAo+SubAoS	-
20	Bitar FF	1998	2m	TF	-

AVC - atrioventricularis comunis; VSD- ventricular septal defect; CoAo- coarctation of the aorta; AoS- aortic stenosis; SubAoS- subaortic stenosis; LHH- left heart hypoplasia; MR- mitral regurgitation; m- months; PDA- persistence of the ductus arteriosus; w- weeks; CAT- common arterial truncus; TF- tetralogy of Fallot; y- years.

tion was performed through median sternotomy and cannulation of the two venae cavae, with extracorporeal circulation at 20°C to 25°C of hypothermia. With the aorta clamped, cardioplegic solution was administered to the aortic root at a dose of 20ml/kg of weight. At the same time, the pulmonary arteries were distally occluded to keep the pressure elevated in the pulmonary trunk.

The first patient underwent surgery in 1987, the second in 1988, the third in 1991 and the fourth in 1997.

## Results

On cardiovascular examination, in addition to the classical signs of CHF, precordial bulging was noted in 3 infants, as well as systolic impulses at the left sternal border and ictus cordis shifted from the midclavicular line in all of them. The cardiac sounds were normal in all infants and a smooth and mild systolic murmur was evident in the mitral area in two patients.

Hepatomegaly was evident and varied from 2 to 4cm from the right costal margin.

Heart rate was increased in all patients and ranged from 120 to 160bpm.

Blood pressure was normal in all infants (mean = 90/60mmHg). There was no edema.

On chest X-ray, cardiomegaly was evident, varying from moderate to marked, and the pulmonary vascular marks were increased due to congestion in all patients.

ECG showed sinus rhythm, with deviation of the electrical QRS axis to the left in all infants, corresponding to +80°, +60°, -40°, and -20°, with necrosis and ischemia in the anterolateral wall, represented by deep and enlarged Q

waves and negative T waves in D<sub>1</sub>, aVL, V<sub>4</sub> to V<sub>6</sub>. Prominent R waves in the left precordial leads indicated left ventricle hypertrophy (LV).

Echocardiographic study confirmed the severe left ventricular dysfunction. The shortening fraction of the myocardial fiber varied from 9% to 23%, with dilation of the left cavities and subvalvar mitral fibrosis. AOLCARPA was accurately diagnosed in 2 out of the 4 patients, and in no patients were anomalous intramural aortic trajectories suspected. In regard to hemodynamic and angiographic studies, they proved accurate in the diagnosis of the anomalous origin of the coronary artery from the pulmonary arterial tree, but they were not useful in the characterization of the peculiar intramural aortic trajectory and also of the origin from the RPA (table II).

In 3 patients, dissection of the left coronary artery during surgery showed that the origin of that artery was in a high position, directly from the RPA. The coronary artery passed behind the pulmonary trunk and entered the aortic wall, running parallel to this vessel's lumen and spreading normally through the walls of the LV. Through the opening of the ascending aorta, the vertical intramural course was clearly visualized. A direct side-to-side anastomosis was performed by linking the intima of the coronary artery with the intima of the ascending aorta to avoid dissection of the latter. Then, ligation of the coronary artery was performed in the RPA. These 3 infants survived for a long-term period that varied from 9 months to 10 years in functional class I and without any medication. There was involution of the electrocardiographic, radiographic and echocardiographic abnormalities to levels considered normal (table III).

In the fourth patient, whose origin of the coronary

**Table II - Results of laboratory tests of anomalous origin of the left coronary artery from the right pulmonary artery with intramural aortic trajectory**

	Clinical Suspicion		Electrocardiogram		Cardiomegaly	Echocardiogram			Angiography
	Coronary anomaly	Intramural course	AQRS	Diagnosis		EF	ΔD	Diagnosis	Potential diagn. of the anom. origin of the left coron. from the RPA
1	Yes	No	+80°	Antlat MI+LVH	Marked	41	17	Dilated cardiomyopathy	Yes
2	Yes	No	-40°	Ant MI STelev.	Marked	55	23	Dilated cardiomyopathy	Yes
3	Yes	No	+60°	Antlat MI	Marked	38	15	Anomalous coron	Yes
4	Yes	No	-20°	Antlat MI + LVH	Marked	20	9	Anomalous coron	-

Antlat - anterolateral; RPA - right pulmonary artery; AQRS - electrical axis of the QRS complex; Coron - coronary artery; ΔD - shortening fraction of the myocardial fiber; diagn - diagnosis; EF - ejection fraction; MI - myocardial infarction; LVH - left ventricle hypertrophy; ST elev. - elevation of the ST segment.

artery was in the bifurcation of the pulmonary trunk with the RPA, a direct end-to-side anastomosis of the left coronary artery with the aorta was performed. This patient developed, in the early postoperative period, a syndrome of low output, requiring intensive inotropic support and peritoneal dialysis for treatment of acute renal failure. Hospital discharge occurred on the 40<sup>th</sup> day of the postoperative period. The infant evolved to functional class II, using digoxin, furosemide and captopril until the 6<sup>th</sup> month, when the patient suddenly died.

### Discussion

AOLCAPA is rarely associated with other cardiac defects, except when the origin is the RPA, a very rare anatomical situation. Of the 20 cases of AOLCARPA reported in the literature <sup>3-21</sup> (table I), 14 had associated defects <sup>3-5,8-10,13,14,16,18-21</sup>, half of which were coarctation of the aorta <sup>5,8,10,13,20</sup>. Interventricular septal defect <sup>4,14</sup> and tetralogy of Fallot <sup>3,21</sup> were also observed. In the 4 cases of AOLCARPA reported in this study, we did not find associated defects but we did find an intramural aortic coronary course, whose diagnosis was established during surgery.

These anatomical peculiarities do not modify the functional alteration usually observed in the anomalous origin of the left coronary artery from the pulmonary trunk. Its main manifestation is represented by ischemic cardiomyopathy, and its evolution essentially depends on the degree of anterograde coronary perfusion from the pulmonary artery or retrograde coronary perfusion from the right coronary artery.

The flow is anterograde, from the pulmonary artery to the anomalous coronary artery, during fetal life and the neonatal period, when pulmonary vascular resistance is still elevated. The flow turns into retrograde, from another coronary artery, after the decrease in pulmonary arterial pressure. From then on, collateral circulation has a significant importance in determining the clinical and evolutionary findings. Myocardial ischemia occurs in situations of inadequate collateral circulation.

Clinical diagnosis of anomalous origin of the left coronary artery should be suspected in young infants with symptoms of CHF and signs of LV cardiomyopathy. In the majority of the patients, there is electrical evidence of ischemia and necrosis on the classic ECG, but the definite diagnosis should be confirmed by echocardiogram, in a

**Table III - Anatomical, surgical and evolutionary characteristics of anomalous origin of the left coronary artery from the right pulmonary artery with intramural aortic trajectory**

	Surgical technique	Age Date	Postoperative evolution							
			FC	Time	Murmur	Liver	ECG	X-ray	SF	EF
1	Direct side-to side anastomosis	5m 6.10.87	I	10 y	-	-	n1	n1	35	70
2	Direct side-to side anastomosis	15m 4.22.88	I	9 y	+ AM	1cm	ST elevation	+	28	61
3	Direct side-to side anastomosis	6m 8.15.91	I	6 y	-	-	n1	n1	36	72
4	Direct end-to-side anastomosis	2m 3.5.97	Death	6m	-	3 cm	LVO antlat MI	++	-	-

MA - mitral area; ant - anterior; FC - functional class; ECG - electrocardiogram; SF - shortening fraction of the myocardial fiber; EF - ejection fraction; MI - myocardial infarction; lat - lateral; m - months; n1 - normal; LVO - left ventricle overload.

conjunct analysis. The most accurate diagnosis requires cardiac catheterization.

Sites of anomalous origin of the coronary artery from the pulmonary arterial tree vary and may include the right sinus, the left sinus, and yet the anterior and posterior sinuses, depending on the position of the pulmonary trunk<sup>17</sup>. In addition to these more usual positions, the origin may be the left pulmonary artery itself<sup>15</sup> and, as happened in our 4 cases, the origin may be high in the RPA or even in the bifurcation of the pulmonary trunk with this artery.

In the direct origin from the RPA, the left coronary artery often follows a vertical course behind the ascending aorta in an inferior direction, for an extension of about 2 cm, passing then close to the posterior portion of the pulmonary ring, and spreading normally through the LV. Although the actual incidence of this peculiar anatom is not known, the descending course of the coronary artery, observed by us, occurs in the posterior wall of the ascending aorta, in the tunica media, emerging from the aorta near the left sinus – hence the difficulty for the echocardiographic diagnosis – passing then behind the pulmonary ring, before the usual distribution through the LV.

In our experience, the echocardiogram also failed to show the intramural aortic trajectory of this anomaly. This may be explained by the close relation between the aorta and the left coronary artery and by the vertical and especially parallel course of this artery. This renders difficult its characterization, unlike the oblique or transverse trajectory in cases of intramural coronary anomalies, such as transposition of the great arteries.

Cardiac catheterization is the most sensitive method for the definitive diagnosis of anomalous origin of the left coronary artery. Aortography points out a dilated right coronary artery with subsequent filling of the anomalous left coronary artery through interarterial collateral vessels, with slight opacification of the pulmonary arterial tree.

However, the intramural aortic trajectory of the coronary artery was not diagnosed in any of our patients in the preoperative period.

Nevertheless, retrospective and careful analysis of the angiography shows some points that may suggest the presence of an intramural aortic trajectory (fig. 1), such as: 1) high origin of the left coronary artery from the pulmonary arterial tree; 2) ascending and parallel course of the anomalous left coronary artery between the origin in the pulmonary artery and the aortic ring; 3) presence of a left right angle formed by the junction of the vertical intramural aortic trajectory and its subsequent horizontal portion until reaching the LV; 4) superoinferior relation between the anomalous ostium of the left coronary artery in the RPA and the ostium of the right coronary artery in the aorta.

In 1988, we published a report of a case of anomalous origin of the left coronary artery from the RPA with an intramural aortic trajectory, as an isolated defect. This was the first report of such a case in the literature<sup>12</sup>. The diagnosis was made during the surgical correction. In 1991, we

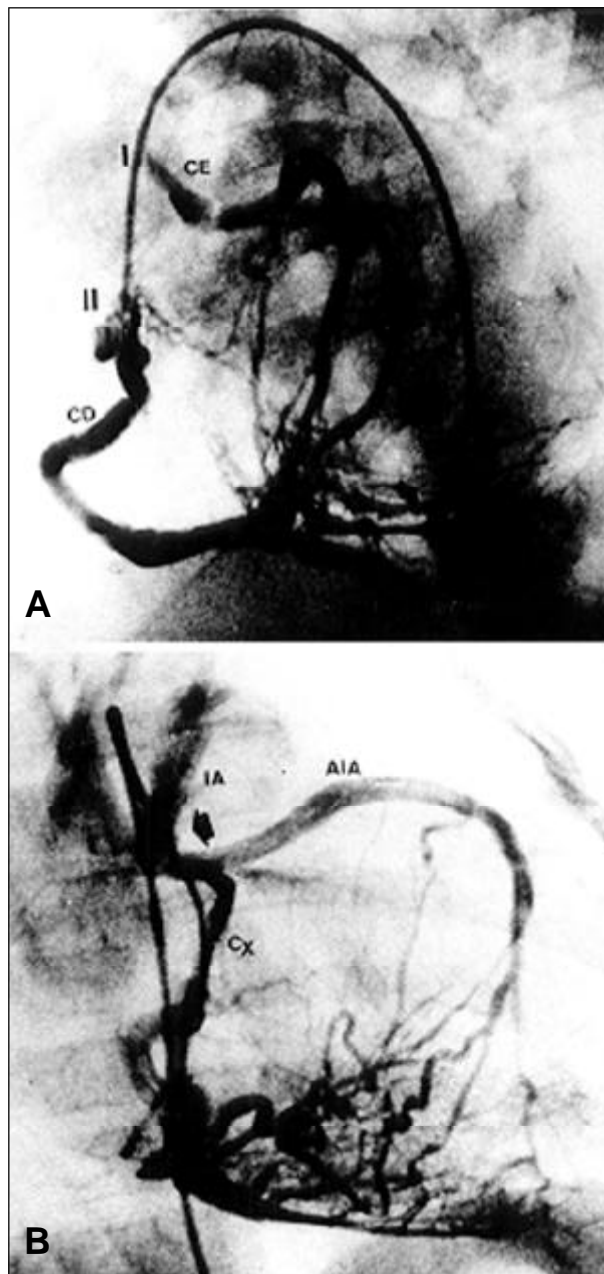


Fig. 1 – Coronary angiography in A (filling of the left coronary artery (LC) by collaterals from the right coronary artery (RC), in left anterior oblique view) and in B (LC in right anterior oblique view), showing the characteristic images of LC origin from the right pulmonary artery: 1) superoinferior relation of the ostium of the LC (I) and of the RC (II), respectively; 2) right angle (arrow) formed in the LC by the vertical (intramural aortic-IA) and the horizontal portion before the bifurcation into anterior interventricular artery (AIA) and circumflex coronary artery (CX); 3) ascending and parallel vertical trajectory of the LC corresponding to the intramural aortic portion; 4) high origin of the LC (A) in relation to the origin of the RC (B).

published the second report of this rare anomaly<sup>15</sup>, using the same surgical technique as in the first case, i.e., direct side-to-side anastomosis of the structures, with success. In 1995, Turley et al<sup>17</sup> published another similar case, and the same surgical procedure was employed. From 1991 to 1997, two other patients with this same anomaly underwent surgery in our service.

Accurate characterization of the vertical course of

the anomalous left coronary artery in the posterior wall of the ascending aorta should be performed through the introduction of an explorer of 1 millimeter of intraostial diameter. With the ascending aorta opened, mobilization of the explorer allows identification of the intramural aortic course that is intraluminally connected, throughout the whole extension of the aorta, creating a large aortocoronary window. A very important subsequent step is the union of the 2 vascular intimal layers, sutured with sepa-

rated stitches to avoid dissection of the aortic wall, as well as occlusion of the coronary lumen.

In our experience, this technique provides simple and safe management of this anomaly with the creation of a large intraluminal aortocoronary window. Usually, the postoperative evolution is good<sup>22,23</sup>, and here we include our 3 patients who underwent this surgical technique and who survived with normalization of the clinical parameters and ventricular function.

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