

## Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery

JORGE O. BARRETTA<sup>1</sup>

The anomaly of the origin of the left coronary artery arising from the pulmonary artery (ALCAPA), was described for the first time in 1885 by Brooks, (1) it is an infrequent and non-lethal congenital heart defect (1 out of 3000 alive newborns) (90% of mortality in the first year of life).

In 1993, Bland et al. were the first in suggest it as a probable clinical diagnosis when facing an infant with signs of left ventricular dysfunction, growth delay, dyspnea, tachycardia, paleness, perspiration and post prandial crying. This disorder is known as Bland-White-Garland syndrome. (2)

ALCAPA has a low incidence and it is accompanied by other structural pathologies. However, it has been described together with several malformations: tetralogy of Fallot, interventricular communication, transposition of the great vessels, (3) coarctation of the aorta (4) and patent ductus arteriosus. Its singular behaviour according to the medical clinic and moment of onset is directly related to the physiopathological changes which accompany the fetus, newborn and infant. The disorder is not verified before birth due to the favorable fetal circulation: high pressure in the pulmonary artery (ALCAPA presents exposure to systemic vascular resistance) and its high oxygen content.

At birth, the presence of patent ductus arteriosus and high level of pulmonary vascular resistances (PVR) expose the left coronary artery to pressure pattern and adequate saturations, but with the consequent ductal closure the oxygen amount in the pulmonary artery decreases. This regular physiological event warns the tributary myocardial territory and generates compensatory mechanisms for flow redistribution, stimulating the intercoronary collateral circulation (ICC).

The progressive and natural decrease of the PVR favor the pressure lowering in the pulmonary artery, with a marked decrease in the perfusion flow, therefore the myocardium will present different degrees of ischaemia depending on the magnitude of ICC development.

The left ventricular anterolateral wall and the homonymous mitral papillary muscle are the most affected parts, two of the elements in which the clinical form of presentation is based (left ventricular dysfunction and mitral regurgitation).

When pressure in the pulmonary artery is low, a steal of coronary flow from the pulmonary artery is added as a last mechanism (*shunt* from left to right).

What initially is myocardial ischaemia will be worsened due to those states that increase the oxygen demand, as diet, crying, pain, fever; giving place to a production of myocardial infarction.

The final product is a manifestation of congestive cardiac failure.

The diagnosis is based on a compatible clinical manifestation, chest x-ray with the presence of cardiomegaly and passive congestion in both lung fields, a characteristic ECG and a Doppler echocardiography. There should be a special mention regarding cardiac catheterization, since the diagnosis should be reaffirmed and at the same time it should be very useful for the surgeon. He should localize the *ostium* of the anomalous artery. With this aim, catheterization should include a pulmonary angiography.

With the advent of new diagnostic imaging techniques (NMR or multislide CAT), today we may argue that the use of these methods is favorable.

It is fundamental, for an early treatment, to spread a bit more "the suspicion of this entity" and consider it as an obligatory differential diagnosis when facing infants with this form of presentation (dilated cardiomyopathy, congenital mitral regurgitation).

Once the diagnosis is made, patients should undergo the surgery with no delay.

The objective should always be to reestablish a dual coronary circulation. (5)

The ALCAPA ligation has been described as a therapeutic option with the aim of interrupting the steal of coronary flow from the pulmonary artery and increasing perfusion pressure in the coronary territory, and although this leads to an improvement in the ventricular function and to a decrease in mitral regurgitation, the risk for sudden death in mid and short-term does not decrease.

The proposed techniques which offer a systemic flow to the left coronary artery (LCA) are diverse, including the *bypass*, but undoubtedly the ideal treatment is the coronary button transfer to the aorta, that is, coronary reimplantation.

The diversity as regards location of the left coronary artery in the pulmonary artery and its branches gave place to surgical strategies, as the one described by Takeuchi et al. in 1979, (6) which consists in tunnel the ostium of the anomalous coronary towards an aortopulmonary window. When the origin of ALCAPA is far from the aorta, for example, in

<sup>1</sup> Main doctor of the Service of Cardiovascular Surgery from Hospital de Pediatría "Prof. Dr. Juan P. Garrahan"

the left pulmonary branch of the pulmonary artery, extensions from this one can be done using the wall of the pulmonary trunk.

In this number of the Argentine Journal of Cardiology, Lugones et al. (7) show an interesting experience in the treatment of this malformation. The surgical results, with the use of the coronary reimplantation technique, have shown that it is a safe procedure with low mortality. Authors as Neirotti (1991), Alexi (1992) and Turley (1995) (8-10) showed in detail a population of patients similar to these ones. In our experience, the use of this method showed that circulatory arrest is not necessary for the correction of this anomaly, even in young and decompensated infants.

The recovery of the ventricular function, even in damaged myocardia, is the rule. An aggressive and early surgical approach is justified, since it represents the only viable possibility to save the hibernating myocardium. The mitral valve improves its function in a high percentage that is why routine mitral plastics should not be done. When facing the lack of improvement after reimplantation or a backward movement in the function during monitoring is urgent to investigate the permeability of the coronary artery through a coronary catheterization.

The best monitoring method in patients with reimplantation of anomalous left coronary artery is controversial, mainly in asymptomatic patients. ECG, effort test and gamma camera have shown contradictory results. (13) That is, there are normal studies even in patients that present an obstruction in the reimplanted coronary arteries. The situation in these patients would be similar to the one in patients who underwent a ligation procedure.

Within monitoring, it is important to confirm the permeability of the implanted coronary artery, since, in presence of obstruction, patients would be exposed to arrhythmias, angina pectoris or sudden death. That is the reason why, angiography or other imaging studies (angioresonance or multislice CT) would have a relevant value.

To the present day, cardiac catheterization has been the gold standard to define the coronary tree anatomy. The indication and the appropriate moment to do a diagnostic catheterization in asymptomatic patients,

as well as the behaviour to follow when facing findings of an occluded coronary artery, are "the discussion".

## BIBLIOGRAPHY

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 cirroid dilatation of the vessels. *J Anat Physiol* 1885;20:26-9.
2. Bland EF, White PD, Garland J. Congenital anomalies of the coronary arteries: report of an unusual case associated with cardiac hypertrophy. *Am Heart J* 1933;8:787-801.
3. Yamagishi M, Emmoto T, Wada Y, Oka T. Transposition of the great arteries with anomalous origin of the left coronary artery from the pulmonary artery. *Ann Thorac Surg* 1998;66:1416-8.
4. Neufeld HN, Schneeweiss A. *Coronary Artery Disease in Infants and Children*. Philadelphia: Lea & Febiger; 1983.
5. Bunton R, Jonas RA, Lang P, Rein AJ, Castaneda AR. Anomalous origin of left coronary artery from pulmonary artery. Ligation versus establishment of a two coronary artery system. *J Thorac Cardiovasc Surg* 1987;93:103-8.
6. Takeuchi S, Imamura H, Katsumoto K, Hayashi I, Katohgi T, Yozu R, et al. New surgical method for repair of anomalous left coronary artery from pulmonary artery. *J Thorac Cardiovasc Surg* 1979;78:7-11.
7. Lugones I, Kreutzer C, Román MI, Schlichter AJ. Origen anómalo de la coronaria izquierda en la arteria pulmonar: resultados de la cirugía correctora. *Rev Argent Cardiol* 2010;78:411-6.
8. Neirotti R, Niyveld A, Ithuralde M, Quaglio M, Seara C, Lubbers L, et al. Anomalous origin of the left coronary artery from the pulmonary artery: repair by aortic reimplantation. *Eur J Cardiothorac Surg* 1991;5:368-71.
9. Alexi-Meskishvili V, Hetzer R, Weng Y, Lange PE, Jin Z, Berger F, et al. Anomalous origin of the left coronary artery from the pulmonary artery. Early results with direct aortic reimplantation. *J Thorac Cardiovasc Surg* 1994;108:354-62.
10. Turley K, Szarnicki RJ, Flachsbarth KD, Richter RC, Popper RW, Tarnoff H. Aortic implantation is possible in all cases of anomalous origin of the left coronary artery from the pulmonary artery. *Ann Thorac Surg* 1995;60:84-9.
11. Laks H, Ardehali A, Grant PW, Allada V. Aortic implantation of anomalous left coronary artery. An improved surgical approach. *J Thorac Cardiovasc Surg* 1995;109:519-23.
12. Del Nido PJ, Duncan BW, Mayer JE, Jr, Wessel DL, LaPierre RA, Jonas RA. Left ventricular assist device improves survival in children with left ventricular dysfunction after repair of anomalous origin of the left coronary artery from the pulmonary artery. *Ann Thorac Surg* 1999;67:169-72.
13. Villafañe, Molina, Salgado, Lopez, Sciegata, Capelli, Laura. Anomalous left coronary artery from the pulmonary artery: mild and long term result after surgery. The fourth World Congress of Pediatric Cardiology and Cardiac Surgery. Argentina. Abstract: 22 de septiembre de 2005.