

Coronary artery bypass grafting as a treatment strategy for ALCAPA syndrome: case report

Cirugía de revascularización coronaria como estrategia de tratamiento para el síndrome de ALCAPA: reporte de caso

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ABSTRACT

Coronary artery anomalies include several congenital conditions characterized by abnormal origin or course of any of the three main epicardial coronary arteries. Anomalous origin of a coronary artery from the pulmonary artery is a rare congenital cardiac anomaly. Out of them, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is the most common. We report the case of a patient with ALCAPA syndrome treated with coronary artery bypass grafting.

Keywords: anomalous left coronary artery from the pulmonary artery (ALCAPA), coronary artery anomalies, coronary artery bypass grafting, pulmonary artery.

Abbreviations:

ALCAPA = Anomalous origin of the Left Coronary Artery from the Pulmonary Artery
ALCARPA = Anomalous origin of the Left Coronary Artery from the junction of the main and the Right Pulmonary Artery
AOCAPA = Anomalous Origin of the Coronary Artery from the Pulmonary Artery
ARCAPA = Anomalous origin of the RCA from the Pulmonary Artery
CABG = Coronary Artery Bypass Grafting
CCTA = Coronary Computed Tomography Angiography
CMR = Cardiac Magnetic Resonance

RESUMEN

Las anomalías de las arterias coronarias incluyen varias afecciones congénitas caracterizadas por un origen o curso anormal de cualquiera de las tres arterias coronarias epicárdicas principales. El origen anómalo de una arteria coronaria a partir de la arteria pulmonar es una anomalía cardíaca congénita poco común. De éstas, el origen anómalo de la arteria coronaria izquierda a partir de la arteria pulmonar (ALCAPA, por sus siglas en inglés) es el más común. Presentamos el caso de una paciente con síndrome de ALCAPA tratado mediante revascularización coronaria.

Palabras clave: arteria coronaria izquierda anómala procedente de la arteria pulmonar (ALCAPA), anomalías de las arterias coronarias, injerto de derivación de la arteria coronaria, arteria pulmonar.

ICA = Invasive Coronary Angiography
LIMA = Left Internal Mammary Artery
LMCA = Left Main Coronary Artery
TTE = Transthoracic Echocardiogram

Anomalous origin of the coronary artery from the pulmonary artery (AOCAPA) is a rare congenital cardiac anomaly, with an incidence of 0.01% in

How to cite: Coyote-Armenia EE, Toledo-García S, Alcalá-Gutiérrez DA, Torres-Perales FDJ, González-Enríquez A, de Paz-Ocaña A, et al. Coronary artery bypass grafting as a treatment strategy for ALCAPA syndrome: case report. *Cir Card Mex*. 2025; 10 (3): 82-86. <https://dx.doi.org/10.35366/120488>

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Received: 03-17-2025. Accepted: 04-02-2025.

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the general population. There are three forms of AOCAPA; namely, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is the most common one, accounting for 1 in 300,000 live births; anomalous origin of the RCA from the pulmonary artery (ARCAPA), and anomalous origin of the left coronary artery from the junction of the main and the right pulmonary artery (ALCARPA).¹

Adults with ALCAPA are characterized by extensive and dilated coronary collateral arteries, cardiomegaly, mitral valve regurgitation, and/or reduced left ventricular ejection fraction. Adults with ALCAPA may be discovered at any point in adulthood and is often an incidental finding when symptoms such as angina pectoris prompt an investigation. Common symptoms of adult ALCAPA, when present, include palpitations from dysrhythmias, syncope, dyspnea, chest pain, and fatigue. ALCAPA is a well-known and important cause of sudden cardiac death in adult patients and should be considered in the differential diagnosis of cardiac arrest.²

Various diagnostic techniques can be used to investigate coronary anatomy and to assess the presence of high-risk features. Coronary computed tomography angiography (CCTA) is currently considered the gold standard.

Current guidelines recommend surgery for all ALCAPA patients regardless of age and symptoms due to lifelong risk of ischemia, ventricular dysrhythmias, and sudden cardiac death.^{3,4} The aim of surgery is to restore a dual coronary system through the reimplantation of the anomalous vessel in the ascending aorta. Several techniques have been designed for this purpose: 1) direct translocation of the anomalous artery (coronary button transfer), which is the technique of choice; 2) the Takeuchi procedure can be performed by creating an intrapulmonary tunnel by means of a parietal flap from the pulmonary artery connecting the anomalous ostium to the aorta (when the anomalous ostium is too distant from the aorta); 3) the anomalous coronary artery can be detached from the pulmonary artery and prolonged to reach the aorta through either a tube created joining flaps from the aorta and the pulmonary artery or oblique coronary prolongation techniques; and finally, 4) coronary artery bypass grafting (CABG) may be an alternative.⁵

Early mortality within 30 days is around 10%, and survival at 20 years following ALCAPA repair has been reported in $86 \pm 4\%$.⁶

We present herein the case of a patient with ALCAPA syndrome treated with ligation of the proximal left main coronary artery (LMCA) and CABG to the distal LMCA using left internal mammary artery.

CLINICAL CASE

A 31-year-old female with a history of dyspnea on exertion and edema in both legs was admitted to the hospital. The

physical examination only revealed edema +/+++ in the lower limbs. Laboratory studies and chest X-ray did not show any alterations. An echocardiogram was performed revealing reverse flow in the proximal portion of the pulmonary artery, and coronary fistula was suspected. CCTA concluded an anomalous origin of the left main coronary artery from the pulmonary artery associated with an extensive network of collaterals between the right and left coronary circulation, and ectasia of all coronary arteries (*Figure 1*). The heart team decided to take the patient for surgical correction of the congenital coronary anomaly.

Surgical procedure

The cardiac approach was performed through a median sternotomy. The pericardium was opened. ALCAPA was corroborated. Left internal mammary artery (LIMA) was harvested. Central cannulation on the ascending aorta and right atrium was used for cardiopulmonary bypass. Aortic and pulmonary cross clamps were used, and cardiac arrest was achieved using antegrade cardioplegia through the ascending aorta and proximal pulmonary artery. Ligation of the left main coronary artery at its origin on the pulmonary artery was made using a “U” stitch with 4-0 polypropylene suture reinforced with teflon pledget. An anastomosis was performed between LIMA and the distal portion of the left main coronary artery (just before its bifurcation into the

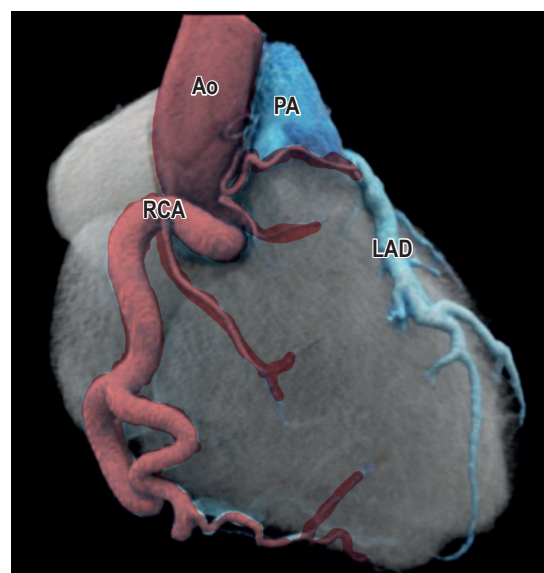


Figure 1: Coronary CT reconstruction showing the right coronary artery with its usual origin from the aorta and the left anterior descending artery with its anomalous origin from the pulmonary artery.

Ao = aorta. LAD = left anterior descending. PA = pulmonary artery. RCA = right coronary artery.

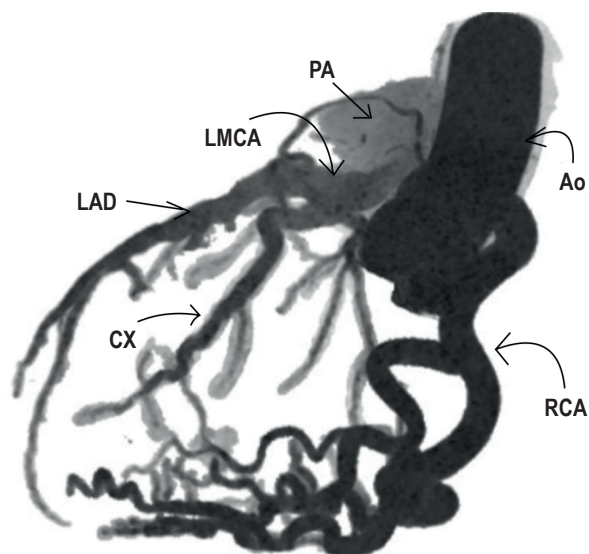


Figure 2: Coronary CT reconstruction showing the right coronary artery with its usual origin from the aorta and the left main coronary artery with an anomalous origin from the pulmonary artery.

Ao = aorta. CX = circumflex. LAD = left anterior descending. LMCA = left main coronary artery. PA = pulmonary artery. RCA = right coronary artery.

left anterior descending and circumflex arteries) using 7-0 polypropylene suture with a parachute technique (*Figures 2 and 3*). Cardiopulmonary bypass was weaned off as per protocol. Cross-clamping and cardiopulmonary bypass time were 93 and 138 minutes, respectively. The patient had a satisfactory postoperative evolution, being extubated within the first six postoperative hours, and discharged from the hospital on the 4th day after surgery. The control electrocardiogram showed no evidence of ischemia. At two months of follow-up, the patient denied angina and improved to New York Heart Association functional class I.

COMMENT

AOCAPA is a rare congenital cardiac anomaly, with an incidence of 0.01% in the general population. As previously mentioned above, three forms of AOCAPA exist: ALCAPA, ARCAPA and ALCAPRA.¹ ALCAPA is the most common type, and was first clinically described in 1933 by Edward Bland, Paul Dudley White, and Joseph Garland, ALCAPA is, therefore, also known as Bland-White-Garland syndrome.⁷

The pathophysiological explanation of ischemia in patients with ALCAPA is as follows: after birth, pulmonary vascular resistance drops, resulting in retrograde flow from the left coronary artery to the pulmonary artery (left to right shunt); this, in turn, leads to reduced coronary perfusion, myocardial ischemia, and resultant ventricular dysfunction as well as other complications including dysrhythmias, mitral regurgitation,

and sudden cardiac death. However, adequate collateral circulatory development can provide adequate coronary perfusion, resulting in a relatively asymptomatic clinical course in a select few, allowing for a late onset presentation, including during adulthood.^{8,9}

ALCAPA is classified into two subtypes by age: those who present in infancy (85%) and, more uncommonly, those who present as older children, teens, or adults (15%), i.e., the so-called adult subtype.² Adult ALCAPA is characterized by extensive and dilated coronary collateral arteries, cardiomegaly, mitral regurgitation, and/or reduced left ventricular ejection fraction. Adult ALCAPA may be discovered at any point in adulthood and is often an incidental finding when symptoms such as angina pectoris prompt an investigation. Common symptoms of adult ALCAPA, when present, include palpitations from dysrhythmias, syncope, dyspnea, chest pain, and fatigue. ALCAPA is a well-known and important cause of sudden cardiac death in adult patients and should be considered in the differential diagnosis of cardiac arrest.² The symptoms reported by our patient, and for which the diagnostic approach was initiated, were dyspnea and edema. There was no angina, palpitations, or syncope. The physical examination only revealed edema +/+++ in the lower limbs.

Various diagnostic techniques can be used to investigate coronary anatomy and to assess the presence of high-risk features. CCTA is currently considered the gold standard.⁵ Electrocardiographic findings suggestive of ALCAPA are abnormal deep or wide Q waves, inverted T waves, and poor R wave progression in leads I, aVL, and precordial lead V4 to V6. Our patient had poor R wave progression on these leads. Transthoracic echocardiogram (TTE) with

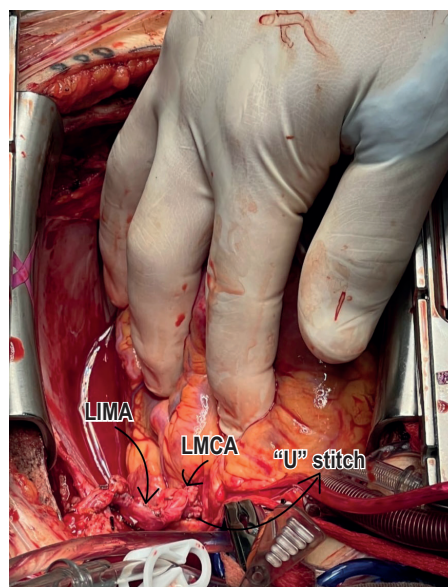


Figure 3:

Left internal mammary artery graft anastomosed to the distal left main coronary artery. A "U" suture was used to close the proximal portion of the left main coronary artery. LIMA = left internal mammary artery. LMCA = left main coronary artery.

color Doppler is a safe, readily available, inexpensive, and portable noninvasive method for initial investigation used in all patients. Echocardiographic findings indicative of ALCAPA include visualization of the left coronary artery originating from the pulmonary artery, retrograde flow from the left coronary artery to the pulmonary artery, dilated and tortuous right coronary artery, lack of the left coronary artery at aortic origin, significant and dilated collateral coronary arteries, mitral regurgitation, left ventricular dysfunction with regional wall motion abnormalities, and enhanced echogenicity of papillary muscles. In our patient, an abnormal reverse flow could be observed in the pulmonary artery just above the pulmonary valve, which was initially misdiagnosed as a probable coronary fistula.² Invasive coronary angiography (ICA) was considered the most important and definite tool to identify and classify CAAs. However, because of its invasiveness, relatively low spatial resolution, and lack of 3-dimensional images, it has been progressively replaced by CCTA.⁵ Typical findings on ICA include a dilated and tortuous right coronary artery with multiple collaterals to the left coronary system; anomalous flow into the pulmonary artery can also be seen. In our patient, ICA was not performed. Some features that can be seen on the CCTA are direct visualization of the left coronary artery originating from the pulmonary artery, a dilated right coronary artery with extensive coronary collateral arteries, abnormal left ventricular wall movement, and dilated bronchial arteries. In infants, the right and left coronary arteries may appear normal. However, the coronary arteries are dilated and tortuous in adults, with dilated intercoronary collateral arteries coursing along the epicardial surface of the heart or within the interventricular septum. In our patient, CCTA allowed us to make the appropriate diagnosis by observing the origin of the left main coronary artery from the pulmonary artery. In addition, there was an important collateral network between the right and left coronary circulation, as well as significant ectasia of the right coronary artery. Cardiac magnetic resonance (CMR) has emerged as an alternative to CCTA in patients with CAAs. In CMR, left ventricular hypertrophy secondary to chronic myocardial hypoperfusion, mitral insufficiency or prolapse, myocardial ischemia, left ventricular wall motion abnormalities, and delayed subendocardial enhancement imply ALCAPA. The retrograde flow from the left coronary artery to the main pulmonary artery, which represents the coronary steal phenomenon can be depicted on CMR.² In our patient, CMR was not performed.

Natural history studies of anomalous coronary artery from the PA (particularly anomalous left coronary artery from the PA) suggest poor outcome in untreated patients.³ Myocardial ischemia is considered the primary cause of life-threatening events in patients with CAAs. Pulmonary

origin of coronary arteries is invariably associated with severe impairment of myocardial perfusion. Although patients with anomalous pulmonary origin of the right coronary artery may be asymptomatic or complain of mild angina or dyspnea, some cases of severe symptoms and even SCD have been reported. On the contrary, anomalous pulmonary origin of the left coronary artery may be rapidly lethal in > 90% of cases, defining the so-called infant type. Notably, a later diagnosis, or adult type, should not be univocally associated with a good prognosis.⁵

Current guidelines recommend surgery for all ALCAPA patients regardless of age and symptoms due to lifelong risk of ischemia, ventricular dysrhythmias, and sudden cardiac death.^{3,4} The aim of surgery is to restore a dual coronary system through the reimplantation of the anomalous vessel in the ascending aorta: direct translocation of the anomalous artery (coronary button transfer) is the technique of choice. When the anomalous ostium is too distant from the aorta, the Takeuchi procedure can be performed by creating an intrapulmonary tunnel by means of a parietal flap from the pulmonary artery connecting the anomalous ostium to the aorta. The anomalous coronary artery can be detached from the pulmonary artery and prolonged to reach the aorta through either a tube created joining flaps from the aorta and the pulmonary artery or oblique coronary prolongation techniques. Finally, coronary artery bypass graft surgery may be an alternative, especially in adults, in whom translocation can be difficult.⁵ Our decision to proceed with revascularization surgery employing the left internal thoracic artery was based on two primary considerations. Firstly, the anatomical configuration of the left main coronary artery, which originated from the non-facing sinus, rendered translocation a technically demanding task. Secondly, our institutional experience with translocation techniques was limited.

Results after surgical repair are excellent. On a single center experience, a total of 78 patients underwent ALCAPA repair. Following repair, systolic LV and MV function improved significantly. Early mortality (within 30 days) was 10% (n = 8). No 30-day mortality was reported in the past 20 years. Survival at 20 years following ALCAPA repair was 86 ± 4%.⁶

CONCLUSION

ALCAPA is the most common form of AOCAPA. In adults (15% of all ALCAPA syndromes) is often an incidental finding when symptoms such as angina pectoris prompt an investigation. CCTA is considered the gold standard diagnostic technique. Current guidelines recommend surgery for all ALCAPA patients regardless of age and symptoms due to lifelong risk of ischemia, ventricular dysrhythmias, and sudden cardiac death. Direct translocation of the anomalous artery (coronary button transfer) is the technique of choice,

but it can be difficult to perform in adult patients, in whom CABG (preferably with an internal thoracic artery graft) is an excellent alternative. To our knowledge, there are no studies that analyze the long-term outcome of adult patients in whom ALCAPA was treated with CABG. We hope that our report contributes to the literature on patients with ALCAPA treated with CABG.

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Funding: none.

Disclosure: the authors have no conflicts of interest to disclose.