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# Association of Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery with Aortopulmonary Window. Two case report

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## Summary

*Anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare entity. Brooks described first cases in 1885. Only 25% to 30% % of cases are associated with congenital defects such as aortopulmonary window and tetralogy of Fallot. It is recommended the reimplantation of the right coronary artery in the Aorta, with redistribution of coronary flow avoiding the signs of ischemia or other complications even when the diagnosis is done in asymptomatic patients. This is the report of two infants with who debuted with murmur and signs of heart failure. ARCAPA and Pulmonary Aortic Window were diagnosed and they were surgically corrected through intrapulmonary tunneling with a favorable evolution.*

**Keywords:** Infant murmur; (ARCAPA) Anomalous Origin of the Right Coronary Artery from Pulmonary Artery.

## Introduction

Anomalies of the origin of the coronary arteries are rare. The anomalous origin of the right coronary artery from the pulmonary artery; (ARCAPA) is a rare entity, which were described in 1885 by Brooks. Many patients are asymptomatic and the diagnosis is made frequently in adults, in the Pathological Anatomy services, so its real prevalence must surely be underestimated [1].

An incidence of 0.002% is reported. In the literature, at least 100 cases of ARCAPA were described and 50% were diagnosed incidentally in the context of an evaluation by heart murmur and signs of ischemia were observed in the electrocardiogram in some patients. Only 25% to 30% of cases are associated with birth defects. Aortopulmonary window (APW) and tetralogy of Fallot are the most frequent. Coronary implantation anomalies are potentially serious entities that can cause sudden death, so surgical correction would be indicated even in asymptomatic patients [1, 2].

Clinical presentation of ARCAPA depends on the degree of collaterals between the right and the left coronary arteries. Most of patients are asymptomatic, however they can rarely present with angina, heart failure, myocardial infarction or sudden death [3-5].

There are no previous reports of this association of heart diseases in our country.

**Clinical Case 1:** One month old female infant, product of a dystocic delivery by caesarean section at 34 weeks gestation (second twin), normal weight. A murmur was auscultated and she was sent to our center for evaluation. A pink, normal weight patient was received with signs of heart failure due to respiratory distress and profuse sweating during suction. On physical examination: polypnea, subcostal and intercostal retraction without rales and a respiratory rate of 69-x minute. Cardiovascular system: rhythmic heart sounds of good tone, systolic murmur in base III / VI, no third noise, second noise with normal pulmonary component and heart rate of 180 beats per minute. Present and synchronous peripheral pulses. Tele cardiogram showing a cardiothoracic index of 0.6 with a predominance of left cavities, pulmonary flow and normal pulmonary artery (PA).

**Electrocardiogram:** Normal axis without alterations.

**Echocardiogram:** Dilation of left cavities, PA and branches. Blind right coronary sinus. Turbulence inside the PA at 11mm from the pulmonary valve plane with diameters of approximately 4mm with, a short circuit from left to right that may be related to pulmonary aortic window or with abnormal origin of the right coronary artery, which indicated the contrasted study (Figure 1).

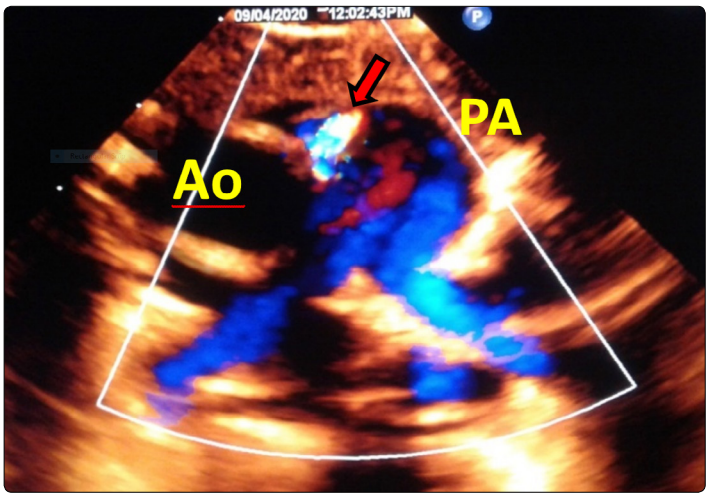


**Figure 1:** Turbulence inside the TAP 11mm from the pulmonary valve plane with a short circuit from left to right. Ao (Aorta), PA (Pulmonary Artery)

**Angiotomography:** Dilation of the left cavities, no aortopulmonary window, predominant left coronary artery, and a suggesting image that could be a right coronary artery emerging from the Pulmonary Artery (Figure 2A).

Hemodynamic and Angiography Report: Pathological saturation differences between the Superior Vena Cava and the Pulmonary Artery. Normal pressure in the pulmonary artery but in upper limit. QP / QS: 4.79. Normal lung resistances. When contrast was injected at the origin of the Left Coronary Artery, progressively opacification of the branches of the Right Coronary Artery was observed, which ended at the Pulmonary Artery.

**Conclusions:** Origin of the right coronary in the Pulmonary Artery (Figure 2B).

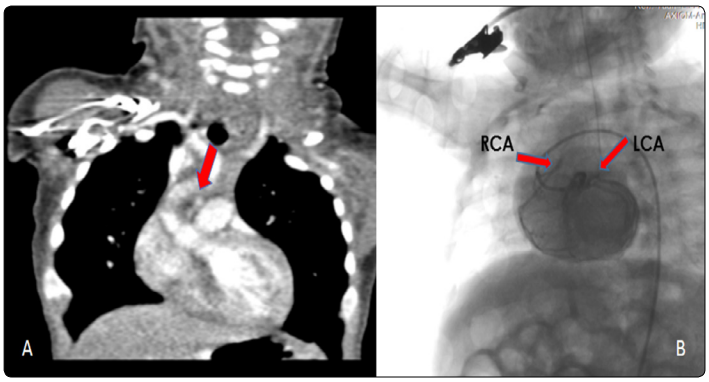


**Figure 2: A:** Image suggestive of abnormal emergence of the right coronary artery. **B:** High origin of the right coronary artery from contrast injection into the left coronary artery. (RCA right coronary artery, LCA left coronary artery)

During surgical operation, ARCAPA was confirmed and it was identified the presence of a diminute APW of 2-3 mm that could justify the elevated QP / QS described by hemodynamics, in an intermediate position and very close to the emergence of the right coronary artery on the anterior wall of the pulmonary artery above the valve plane (Figure 3).

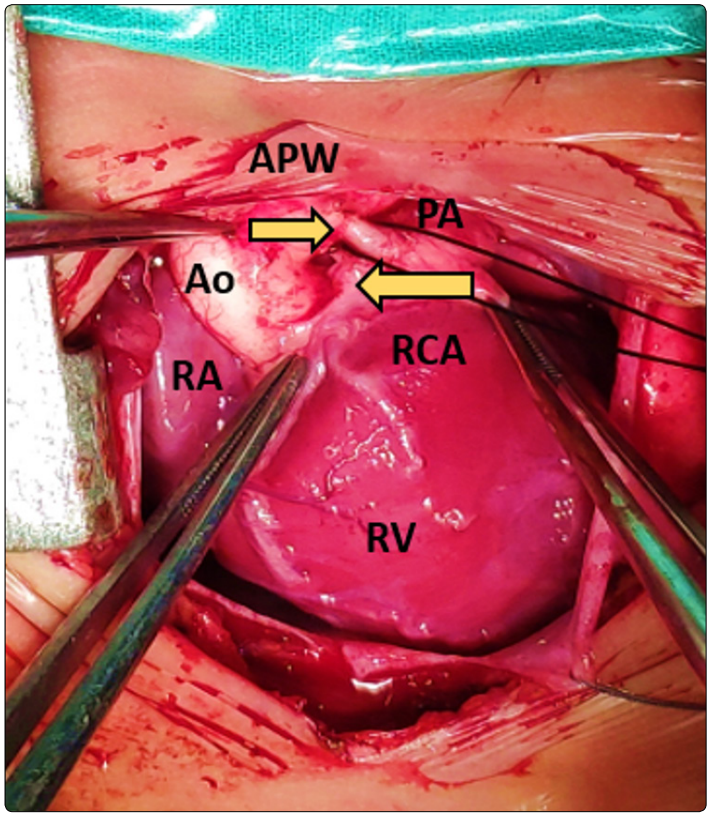


**Figure 3:** APW, aortopulmonary window; PA, pulmonary artery; Ao, aorta; RA, right atrium; RV, right ventricle; RCA, right coronary artery.

**Clinical Case 2:** Two-month-old infant with a health history who, in a regular pediatric consultation, was detected a murmur and signs of heart failure, due to respiratory distress, limited sucking and sweating. He was referred to our center for study and treatment. Physical examination of the respiratory system found polypnea, subcostal and intercostal retraction, discreet nasal flutter, isolated fine wheezing in both lung fields. Respiratory rate was 80 x minute.

**Cardiovascular System:** Rhythmic heart sounds of good tone, continuous murmur in base III / VI, no third noise, second component noise reinforced, and heart rate 170-x minute. Peripheral pulses present and normal. Tele cardiogram showed a cardiothoracic index of 0.64 with a predominance of left cavities, increased pulmonary flow and rectified PA.

**Laboratory Tests:** Blood count, blood chemistry and coagulogram within normal limits.

**Electrocardiogram:** Normal axis with no signs of ischemia or other abnormalities. Echocardiogram: Aortopulmonary window with hemodynamic repercussion.

Performing the corrective surgery of the APW, the anomalous emergency of the right coronary was detected (Figure 4A). Through the pulmonary arteriotomy, the right coronary ostium was located in the anterolateral wall of the pulmonary artery in the right posterior facing sinus below the lower border of the APW (Figure 4B).

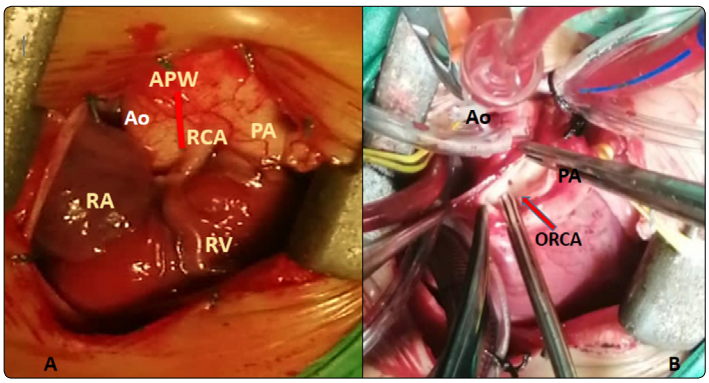


**Figure 4: A:** red line: external view of APW, aortopulmonary window; PA, pulmonary artery; Ao, aorta; RA, right atrium; RV, right ventricle; RCA, right coronary artery. **B:** ORCA, ostium of right coronary artery

In both patients, an autologous pericardium patch was placed through the pulmonary artery from the upper edge of the aortopulmonary window to a position below the emergence of the right coronary artery, in the first case above the pulmonary valve plane and in the second, within the sinus of the pulmonary sigmoid, allowing anterograde coronary filling through the native window. There were no a residual short circuit, an obstructive gradient in the right outlet or a postoperative electrocardiographic alteration reported.

## Discussion

Congenital anomalies of the coronary arteries are a set of vascular anatomical variants present from birth. ARCAPA is an infrequent coronary anomaly, it can be present in structurally normal hearts in 30%, associated with other congenital heart diseases, Tetralogy of Fallot and the APW are the most frequent [2]. At least 100 cases of ARCAPA have been described, including pediatric and adult patients with several clinical manifestations. Most were incidentally diagnosed in the context of an evaluation for a heart murmur. There are differences from the debut of the anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), which appears an early age with signs of anterolateral myocardial infarction, in most the patients [2, 6].

This congenital heart disease can also develop symptomatically, due to the phenomenon of coronary steal that occurs from the left coronary artery to the right through collateral vessels. In this case, patients may present with symptoms of myocardial ischemia, congestive heart failure, even sudden death. Severity depends on the impact of the abnormality on the delivery of oxygen to the myocardium, the direction of flow, and the formation of collateral circulation [7].

There are protective situations associated with ARCAPA, such as stenosis of the origin of the abnormal right coronary artery, left coronary dominance, and the presence of short circuits from left to right (ventricular defects, persistent ductus arteriosus, or aortopulmonary window) [2].

Neither of the two clinical cases showed collateral circulation, but they did present some of these protective situations, such as the presence of left-to-right short circuits and left dominance in case one. In-patient 1, diagnosis of ARCAPA was made in an infant with signs of heart failure and with a systolic murmur, suggestive echocardiographic image with a left-to-right shunt showed the lesion. Presence the APW detected during surgery could justified the high QP / QS values observed in hemodynamics; the proximity of both lesions in the wall of the pulmonary artery and the dimensions of the window made diagnosis difficult from the beginning.

In patient 2, who was admitted with severe symptoms of heart failure due to the hemodynamic repercussion of the APW, the dilation and increased pressure in the pulmonary artery, might had masked or decreased coronary steal and its expression in suspicious or indirect echocardiographic signs of ARCAPA, that´s why it was a finding in the surgical act.

Despite having arrived with symptoms at the service, they quickly were compensated with medical treatment without elements of myocardial ischemia on the electrocardiogram in any time. Despite the low frequency and the fact that ARCAPA is often diagnosed by pathological anatomy and in adults. Reimplantation of the right coronary artery in the aorta is recommended, with the redistribution of coronary flow avoiding the signs of ischemia or other complications even when the diagnosis is made in asymptomatic patients, on the contrary the results in those who go to surgery without any type of clinical manifestation is to expect a better postoperative and recovery [2, 8-10]. The patients in this report presented a favorable preoperative evolution.

Coronary reimplantation was not performed, but rather a variant of intrapulmonary tunneling, as it seemed a feasible procedure for the solution of both malformations and less risky for our small patients, who showed a satisfactory postoperative evolution. Both cases are included in the small percentage of the coronary syndrome that are accompanied by another congenital heart disease.

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