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An Unusual Combination of an Anomalous Origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA) and a Right Coronary Artery System with Two Separate Ostia from the Aorta in an Adult

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We describe a patient with an infrequent combination of variants in both the right and the left coronary arterial ostia, namely a combination of two separate right coronary artery (RCA) ostia from the aorta, and an anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). To our knowledge, such a combination has not been previously reported. Based on published statistics for individual variants, such a combination is expected to occur approximately once for every 500,000 to one million live births. ALCAPA and dual RCA anatomy was characterized in our patient by echocardiography, conventional angiography, and multidetector computed tomography before and after Takeuchi repair. (Echocardiography 2010;27:E13-E17)

Key words: echocardiography, computed tomography, anomalous coronary artery, myocardial ischemia, congenital heart disease

Case Report:

A 28-year-old healthy previously asymptomatic woman with a history of a childhood murmur and three prior uneventful pregnancies was referred to cardiology clinic for evaluation of a 9-month history of exertional dyspnea. Physical examination revealed a soft, grade 2/6 holosystolic murmur at the apex radiating to the axilla. Electrocardiogram showed sinus rhythm and no evidence of ischemia or infarction.

Transthoracic echocardiography ordered for evaluation of the murmur demonstrated moderate mitral regurgitation with a posteriorly directed jet due to anterior mitral leaflet prolapse and normal left ventricular ejection fraction of 55%. In addition, a very abnormal color Doppler flow was noted on the parasternal short-axis view at the level of the mitral valve. The flow was seen originating in the posterior atrioventricular groove, extending through the inferior and anterior interventricular septum and extending into the right ventricular free wall (Fig. 1).

On transesophageal echocardiography (Fig. 2), a markedly enlarged right coronary artery (RCA) with a diameter 0.90 cm (normal 0.39 ± 0.06 cm)[1] was seen originating from the correct sinus of Valsalva. The flow in this dilated RCA was in the normal antegrade direction. In contrast, an abnormal retrograde flow in the left anterior descending (LAD) that did not extend into the left aortic sinus of Valsalva was noted (Fig. 3). Additionally, an abnormal color Doppler flow into the main pulmonary artery (MPA) occurring throughout the cardiac cycle was visualized (Fig. 4).

On cardiac catheterization, the ostium of the left coronary artery (LCA) could not be located in any of the sinuses of Valsalva. The RCA had two separate ostia of unequal size in the right sinus of Valsalva (Fig. 5). The larger ostium was that of the principal RCA; the vessel was very tortuous and filled the LCA retrogradely via extensive collaterals (Fig. 5A). The smaller ostium gave rise to the conus artery that provided a small degree of collateral blood flow through the circle of Vieussens to the LAD territory (Fig. 5B). There was a step-up saturation at the level of the proximal MPA indicative of a left-to-right shunt whose magnitude (Qp/Qs ratio) was 1.5:1.0.

Multidetector-computed tomographic (MDCT) coronary angiography demonstrated an anomalous origin of the LCA in the inferior-posterior aspect of the MPA diagnostic of ALCAPA (Fig. 6A and 6B). On 3D-rendered MDCT images, the special relationship between the main RCA, conus branch and their collaterals to the LCA can easily be appreciated (Fig. 6C).

She subsequently underwent Takeuchi surgical repair to restore antegrade flow from the ascending

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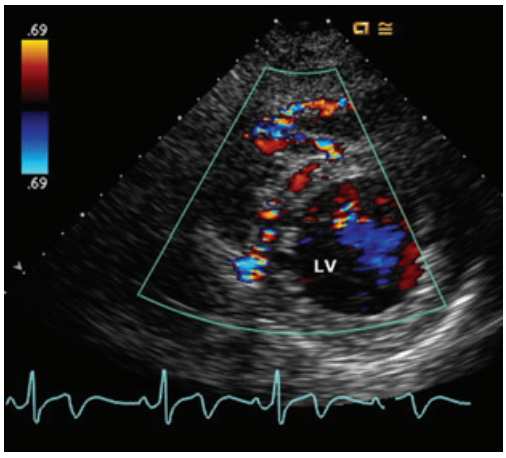

Figure 1. Tranthoracic echocardiogram. Paraternal short-axis view at the level of the mitral valve on transthoracic echocardiography demonstrates an aberrant flow from the right coronary into the branches of the left anterior descending artery and further retrograde into the coronary arteries supplying the right ventricular free wall due to ALCAPA. LV = left ventricle.

aorta into the LCA. The anatomic alterations created by the surgical repair were well seen on repeat MDCT 2 weeks later. The LCA remained attached to the MPA; its ostium was connected to a 3 × 4 mm aortopulmonary window via a baffle (1.4 cm in diameter) across the MPA (Fig. 7). By now the diameter of the ostium of the principal RCA has shrunk to 0.6 cm. A dobutamine stress echocardiogram performed 9 months after surgery was negative for ischemia. The patient remains asymptomatic.

Discussion:

Variations in the branching pattern of coronary arteries are common; however, anomalies of the coronary ostia are rare with a reported incidence of 1–2% in the general population.[2] Many anomalies are benign and are discovered incidentally; the others have been implicated in chest pain, dyspnea, myocardial infarction, left ventricular dysfunction, ventricular fibrillation, and sudden cardiac death.[3]

We describe a patient with an infrequent combination of variants in both the right and the left coronary arterial ostia, namely a combination of two separate RCA ostia from the aorta, and an anomalous origin of the LCA from the pulmonary artery (ALCAPA). The finding of dual RCA ostia in the absence of ALCAPA is very common. It occurs in 33 to 50% of humans and is considered to be a normal variant.[4] However, ALCAPA (also known as Bland-White-Garland syndrome) is very rare and is seen in 1/300,000 live births.[5] To our knowledge, the presence of both ALCAPA and dual RCA ostia in the same patient has not been published previously. Based on

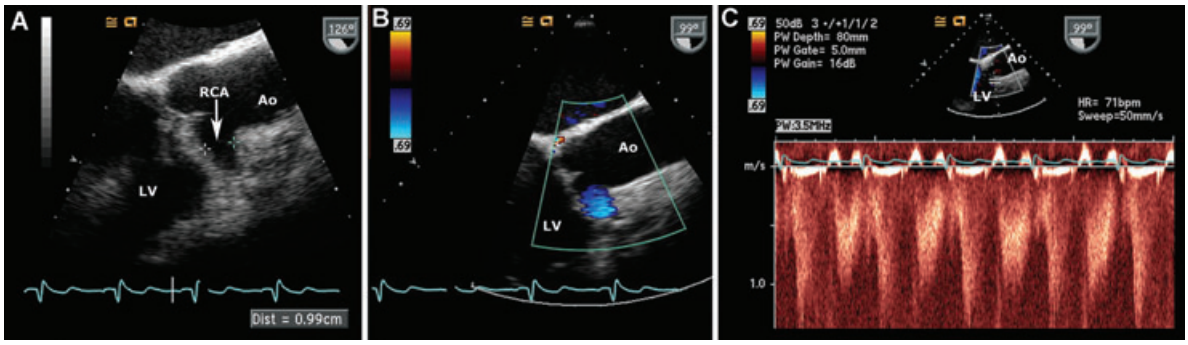

Figure 2. Transesophageal echocardiogram of the RCA. **A.** Markedly dilated ostium of the main RCA seen by gray-scale and **B.** color Doppler imaging. **C.** The spectral Doppler obtained from the right coronary artery reveals an accelerated antegrade flow. Both the RCA dilatation and flow acceleration are consequences of marked right-to-left intracoronary shunting due to ALCAPA. Ao = aorta; LV = left ventricle.

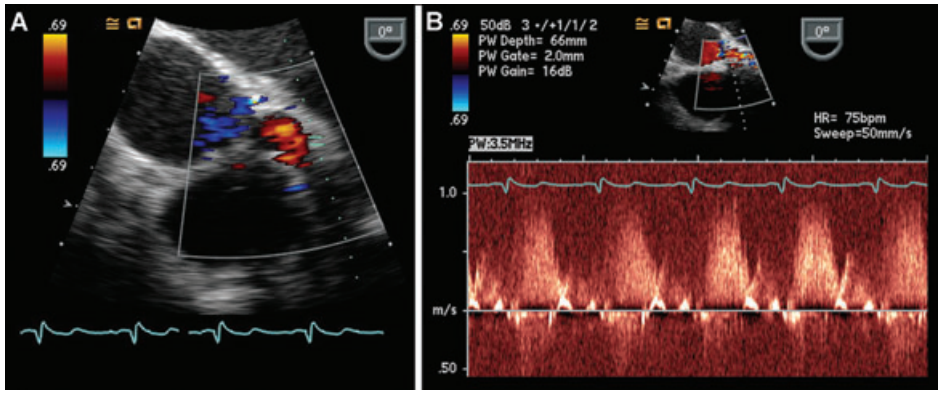

Figure 3. Transesophageal Echocardiogram of the LCA. Flow reversal in the LCA due to ALCAPA is demonstrated by both **A.** color Doppler (arrow), and **B.** spectral Doppler.

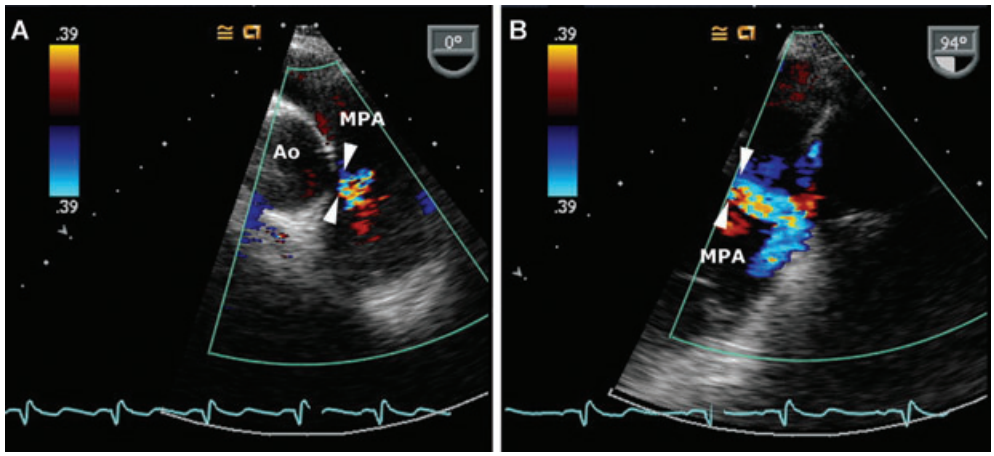

Figure 4. Transesophageal echocardiogram of the MPA. Due to flow reversal in the anomalous left main coronary artery, there is an abnormal color Doppler jet (arrowheads) entering the MPA. This finding is indicative of ALCAPA. Ao = ascending aorta. Imaging was obtained at **A.** 0 degrees and **B.** 94 degrees.

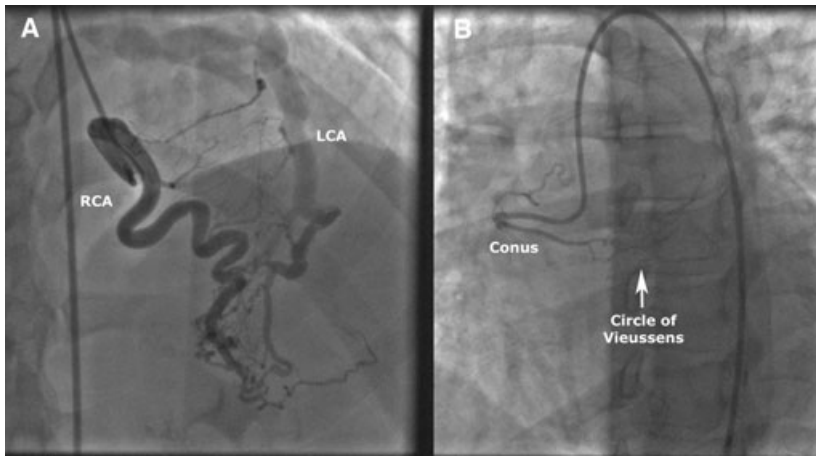

Figure 5. Coronary angiogram. Coronary angiography reveals two separate coronary ostia in the right sinus of Valsalva. **A.** Right anterior oblique view of the aortic root injection shows the origin of the principal RCA that is dilated and tortuous. The contrast is seen flowing antegrade in the principal RCA, then retrograde in the LCA and finally emptying into the presumed main pulmonary artery. **B.** Anteroposterior view with cranial angulation demonstrates the conus artery, which has a separate ostium from the right sinus of Valsalva. It is collateralized to the left anterior descending artery via the circle of Vieussens.

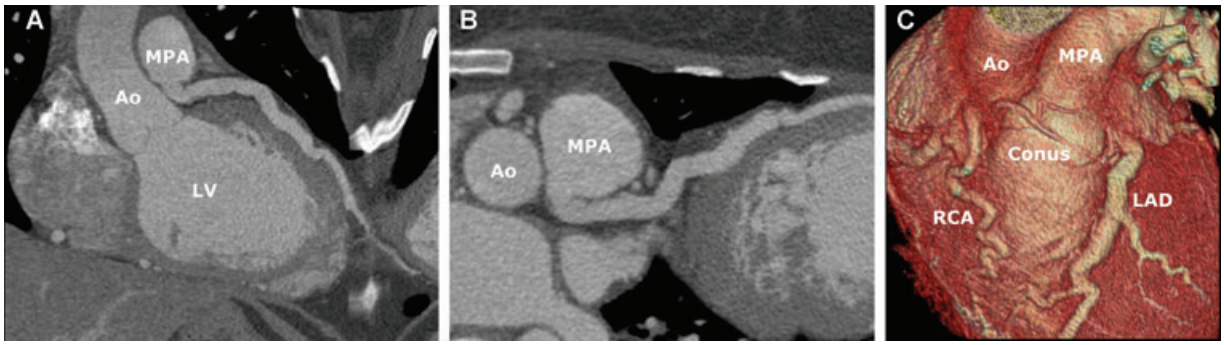

Figure 6. Images from ECG-gated CT coronary angiogram. **A.** Coronal reformatted view shows anomalous origin of the left coronary artery from the main pulmonary artery. **B.** Curved multiplanar reformatted image demonstrates that the origin of left coronary artery is from the posterior aspect of the main pulmonary artery. **C.** Volume-rendered view shows course of LAD and conus branch arising separately from aorta. Ao = aorta; LAD = left anterior descending coronary artery; LV = left ventricle; MPA = main pulmonary artery; RCA = right coronary artery.

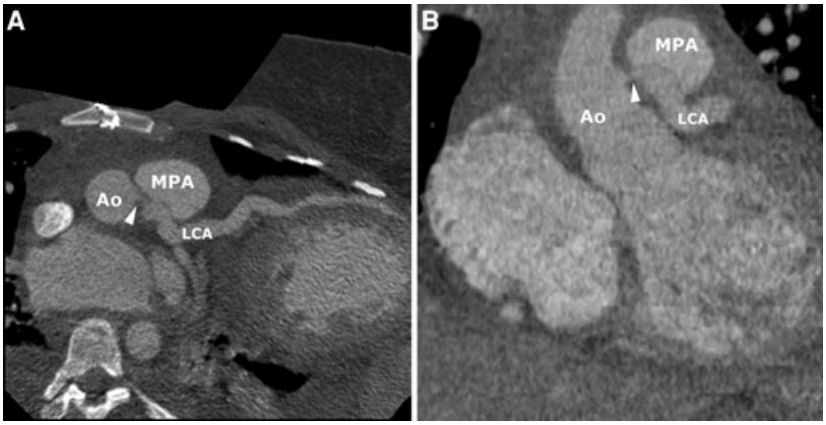

Figure 7. Images from ECG-gated CT coronary angiogram after the Takeuchi repair. **A.** Curved multiplanar reformatted image shows that left coronary artery remains attached to the MPA. The baffle (arrowhead) is seen extending from the aorta into the pulmonary artery separating the origin of the left coronary artery from the pulmonary artery. Ao = aorta; LCA = left coronary artery; MPA = main pulmonary artery.

above statistics for individual variants, such a combination is expected to occur approximately once for every 500,000 to one million live births (Table I).

In ALCAPA patients, systemic and pulmonary pressures are equal in utero that allows for an antegrade flow in both the right and left coronary arteries. In the neonatal period, the pulmonary pressure falls below the systemic pressure and the ductus arteriosus closes. As a result, the direction of flow in the anomalous LCA reverses; the blood from the high-pressure RCA territory artery fills the LCA via extensive collaterals and exits the LCA into the MPA.

Patients who do not develop significant intracoronary collaterals soon after birth are said to have "infantile type" ALCAPA. They present during the first 3 months of life with failure to thrive and may develop myocardial ischemia, left ventricular dysfunction, mitral insufficiency, and death. If untreated, 90% of such children die within the first year of life.[6]

The "adult-type" ALCAPA is rare and represents 10–15% of survivors that have well-established intracoronary collaterals and often no overt myocardial ischemia.[7] The oldest reported patient with unrepaired ALCAPA was 83 years old but this is an exception.[8] Malignant arrhythmias leading to sudden death are common, and 90% of such patients die suddenly at a mean age of 35 years.[6,7] Our patient had no symptoms until the age of 28 years and had three uncomplicated pregnancies and deliveries.

The echocardigraphic diagnosis of ALCAPA requires demonstration of an antegrade flow in an enlarged RCA, the absence of the LCA ostium in the aortic root, and the presence of abnormal retrograde flow in the LCA directed into the MPA.[9] The diagnosis may be supported by conventional coronary angiography or MDCT.

Various surgical techniques of ALCAPA repair have been used including ligation of the anomalous left coronary artery, coronary artery bypass grafting, creation of an intrapulmonary conduit/baffle (Takeuchi procedure) and direct reimplantation of the left coronary artery to the aorta.[10] Operative mortality rate of current techniques ranges from 0 to 23%.[11]

TABLE I

Incidence of Coronary Anomalies

Lesion	Incidence	Reference
Any congenital anomaly of coronary ostia	1–2%	Hoffman et al., 2004[2]
Right coronary artery (RCA) with dual ostia	30–50%	Edwards et al., 1981[4]
Anomalous origin of left coronary artery from pulmonary artery (ALCAPA)	1 per 300,000	Menahem et al., 1987[5]
Expected incidence of both ALCAPA and RCA with dual ostia*	1 to 2 per 1,000,000	Calculated based on the assumption that ALCAPA occurs independent of dual ostia RCA

* If the incidence of dual-ostia RCA (30–50% of births) is independent of ALCAPA incidence (1 in 300,000 births) then the likelihood of both anomalies occurring in the same person is the product of the individual incidences (30–50% × 1/300,000), or approximately 1 to 2 in 1,000,000 births.

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Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clips for Figure 1, Figure 2B, Figure 3A, and Figure 4B.

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