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Anomalous Right Coronary Artery from the Pulmonary Artery: Noninvasive Diagnosis and Serial Evaluation

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ABSTRACT

Isolated anomalous origin of the right coronary artery from the main pulmonary artery (ARCAPA) is a rare congenital cardiac malformation. We reviewed the current literature and found only 31 patients with ARCAPA. We report the first case that was diagnosed and followed on a noninvasive basis with cardiovascular magnetic resonance after surgical re-implantation. This report of a patient with ARCAPA showed resolving coronary artery sizes secondary to decreased pulmonary steal. Cardiovascular magnetic resonance is an accurate and reliable imaging modality that allows serial noninvasive follow up in patients with coronary artery anomalies.

A 39-year-old mother of two presented with a 6 month history of mild left-sided chest pain with exertion. An electrocardiogram demonstrated T-wave inversion. Echocardiography revealed a dilated coronary artery system with a continuous fistulous-type flow and a diastolic jet in the main pulmonary artery. An exercise stress test was normal. A cardiovascular magnetic resonance (CMR) was performed to evaluate the proximal course of the coronary arteries (Figs. 1–3). The CMR showed extremely dilated epicardial coronary arteries. The large, dilated left main coronary artery arose from the left sinus of Valsalva. The left circumflex artery was normal in size, arising appropriately from the left main coronary artery. The entire left anterior descending artery was dilated and tortuous, coursing along the interventricular groove toward the cardiac apex. The right coronary artery was also dilated and tortuous and demonstrated retrograde flow with phase contrast imaging. The origin of the right coronary artery arose from the main pulmonary artery. There was turbulent flow demonstrated with CINE and phase contrast imaging as blood flowed from the right coronary into the pulmonary artery (Fig. 4).

A cardiac catheterization was subsequently performed and confirmed the diagnosis of anomalous right coronary artery from the pulmonary artery (ARCAPA) (Figs. 5 and 6). Surgical correction was accomplished by excising the right coronary artery origin with a button of tissue from the pulmonary artery and re-implanting it to the anterior aspect of the ascending aorta. The postoperative transesophageal echocardiogram showed excellent biventricular function with no segmental wall motion abnormalities. A CMR was performed on postoperative day 6 as a baseline examination for future evaluation. This CMR showed the dilated right coronary artery arising off of the ascending aorta. Antegrade flow was noted although there was evidence of sluggish flow within the dilated and tortuous vessels. She was discharged home without event.

A 6 month postoperative CMR was performed to evaluate the dilated epicardial coronary system. The ostium of the right coronary artery was unchanged in appearance, without evidence of ostial stenosis. The left coronary ostium decreased in size from 8 mm to 6 mm in diameter when compared to the preoperative study. In addition, the proximal left anterior descending artery diameter decreased from 10 mm to 7 mm. The decrease in luminal caliber was likely secondary to decreased coronary flow resulting from eliminating the coronary steal to the pulmonary artery. In addition, there was good biventricular function, without regional wall motion abnormalities.

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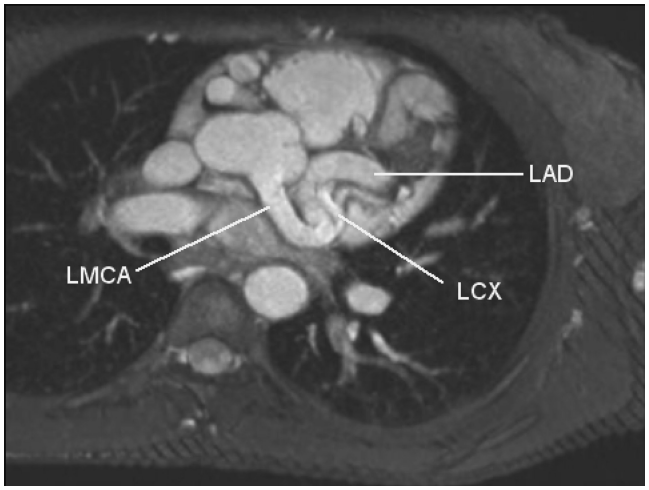

Figure 1. CMR—Left coronary artery. Images from a respiratory-navigator, vectorcardiographic-triggered 3D turbo field echo sequence shows the dilated origin of the left coronary artery. The left main coronary artery (LMCA) and left anterior descending (LAD) are dilated (arrow). The left circumflex artery (LCX) is normal in size.

COMMENT

This report describes the first reported case of ARCAPA definitively diagnosed by CMR and followed on a serial basis. Patients with the anomalous origin of the right coronary artery from the pulmonary artery have numerous collateral vessels directing the blood flow from the left coronary artery through the right coronary artery retrograde into the pulmonary trunk. The continuous turbulent flow seen at the point of entry of the right coronary artery into the pulmonary artery is secondary to a higher flow state into the low resistance pulmonary system. This would explain the continuous fistulous-type flow demonstrated in the pulmonary artery seen by echocardiography. Such reverted flow leads to left-to-right shunt of a borderline hemodynamic significance; however, results in the coronary steal phenomenon with potential consequences of myocardial ischemia. The coronary steal introduces an oxygen delivery/consumption mismatch during physical exertion, causing this patient's symptoms and abnormal electrocardiogram upon presentation. Depending on the amount of coronary steal, patients may or may not have electrocardiographic evidence of ischemia during stress testing. As this anomaly is known to be associated with increased risk of myocardial infarction and sudden death without any preceding symptoms, prophylactic surgical therapy is commonly indicated. After surgical repair, this patient no longer had symptoms of chest pain with exertion, and her ECG showed resolution of the T wave changes.

Coronary artery malformations are rare in the general population with an incidence of only 0.3 to 0.9% (1–3). The

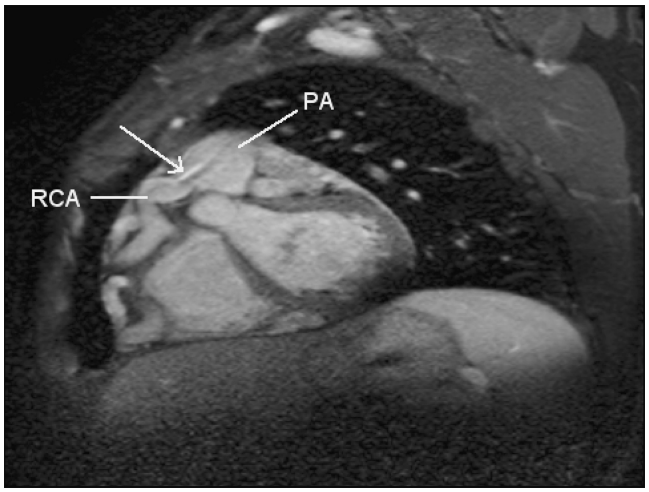

Figure 2. CMR—Right coronary artery. There is marked ectasia of the right coronary artery (RCA) system with dephasing (arrow), indicative of turbulent flow, demonstrated at the origin of the RCA where blood flows into the pulmonary artery (PA).

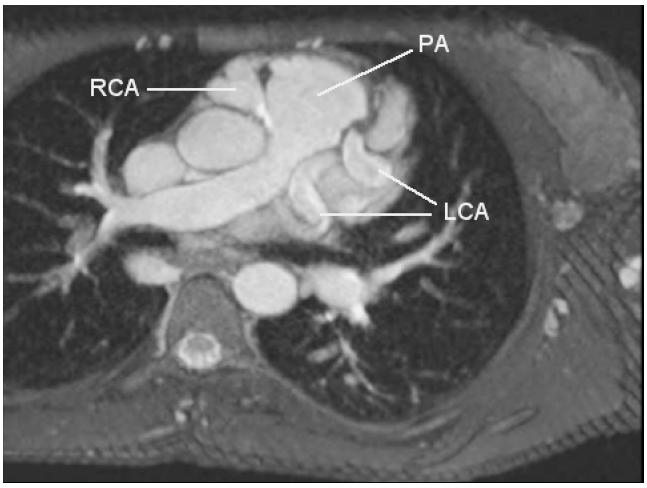

Figure 3. CMR—Tortuous coronary artery system. LCA = left coronary artery, PA = pulmonary artery, RCA = right coronary artery.

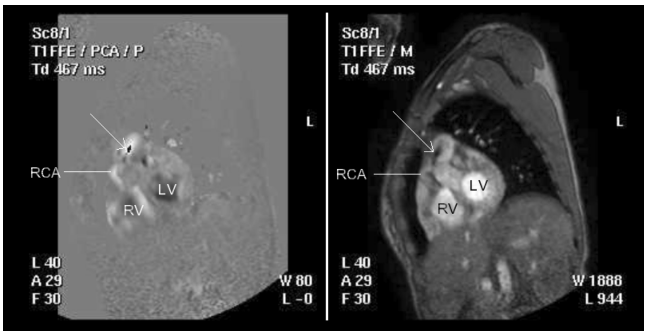

Figure 4. Phase contrast imaging—phase and magnitude images. Dephasing jet (arrow) retrograde from the right coronary artery (RCA) to the pulmonary artery by cine phase contrast sequence. LV = left ventricle, RV = right ventricle.

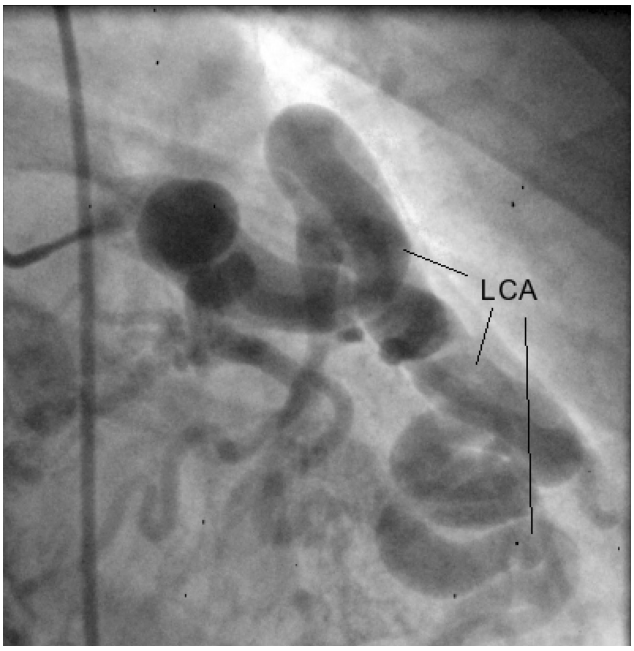

Figure 5. Left coronary artery angiogram. Early filling during the left coronary artery (LCA) angiogram demonstrates a dilated and tortuous coronary system.

incidence of coronary artery abnormalities may be as high as 36% in patients with congenital heart disease (4, 5). Accurate identification of coronary artery anomalies is vital in patients with congenital heart disease, as the pattern and course of the abnormality determines the need for treatment and may affect the type of repair or patient outcome. Coronary artery imaging with echocardiography may be difficult in some patients due to poor acoustic windows. Cardiac computed tomography (CT) is an effective noninvasive test but uses ionizing radiation and requires intravenous administration of iodinated contrast agent. Conventional x-ray cineangiography is an invasive test and may also be difficult because of the lack of 3D information that relates the coronary artery to its surrounding structures (5, 6). Recent advances in CMR have allowed CMR to become a valuable noninvasive test to evaluate coronary arteries without the need for an intravenous contrast agent, radiation exposure, or prolonged breath holding (7). CMR's ability to consistently image coronary arteries underlies the importance of non-invasive imaging as a viable alternative to repeated cardiac catheterization.

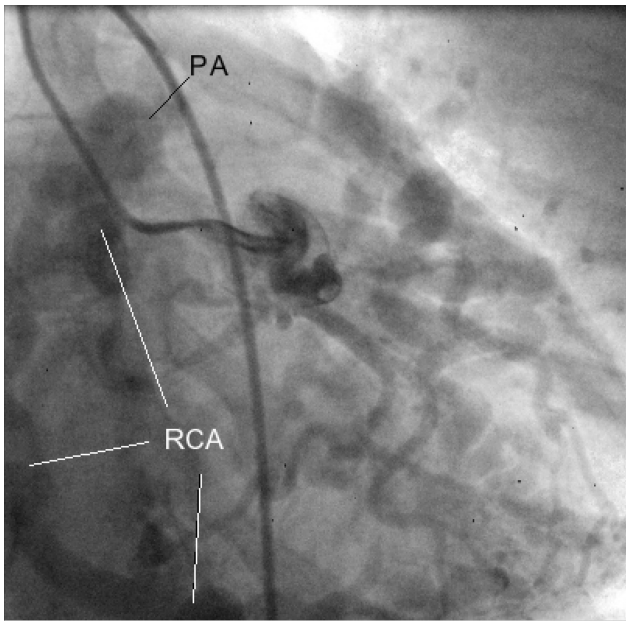

Figure 6. Left coronary artery angiogram—late phase. Later phase after the left coronary artery angiogram demonstrates a dilated right coronary artery (RCA) system that fills via collaterals with retrograde flow demonstrated into the pulmonary artery (PA).

Isolated anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) is a rare congenital cardiac malformation. Unlike anomalous origin of the left coronary artery from the main pulmonary artery, many patients with ARCAPA are often asymptomatic. Given the fact that many patients are indeed asymptomatic, the true prevalence of ARCAPA is likely underestimated. Review of the literature showed only 31 previously reported patients with isolated ARCAPA (Table 1). These patients have a variable clinical presentation, ranging from no symptoms (in 10/26 or 38.5% of patients) to myocardial infarction and sudden cardiac arrest/death (4/26 or 15%). There was also a wide age range at presentation: 2 months to 74 years of age.

In the few cases of isolated ARCAPA that have been reported, surgical repair has been performed at the time of diagnosis, even in the asymptomatic patient. Surgical repair is recommended because there is a risk of an acute event (sudden death or myocardial infarction). In fact, of the 31 patients with isolated ARCAPA, 15% presented with either cardiac arrest or an acute myocardial infarction. The type of surgical repair may include ligation of the anomalous right coronary artery near the pulmonary artery, ligation of the anomalous right coronary artery near the pulmonary artery with a saphenous vein conduit to the aorta, or a button type of excision with reimplantation to the aorta.

CMR has undergone tremendous development over the past decade. This development has included both hardware and software advances, which have allowed for improved noninvasive imaging of the cardiovascular system. The definition of coronary artery origin and course may be difficult with conventional x-ray angiography because images are 2-dimensional projections of a tortuous 3-dimensional structure (5, 6). In addition, the surrounding structures (great arteries, cardiac and mediastinal structures) are not seen well with conventional angiography. With the use of free-breathing respiratory-navigator acquisition CMR, we have the ability to not only visualize the proximal coronary arteries (36), but also determine the blood flow direction and velocity, detect for perfusion abnormalities, and evaluate myocardial viability. CMR is a technique that allows for reliable and accurate visualization of coronary artery abnormalities and also allows these patients to be followed serially with noninvasive imaging.

Table 1. Review of Literature: thirty-one patients with isolated Anomalous Right Coronary from the Pulmonary Artery

Reference	Year reported	Age (yr)	Sex	Symptoms	Diagnosed By	Therapy
Brooks(8)	1886			—	Autopsy	—
Monckeberg (9)	1914	31	M	—	Autopsy	—
Jordan et al. (10)	1950	74	M	—	Autopsy	—
Rhantigan et al. (11)	1971	59	F	—	Autopsy	—
Wald et al. (12)	1971	17	F	Cardiac Arrest	Autopsy	—
Wald et al. (12)	1971	42	F	None	Angiography	Operation
Tingelstad et al. (13)	1972	12	M	None	Angiography	Operation
Eugster and Olivia (14)	1973	64	F	AF, CHF	Angiography	Operation
Bregman et al. (15)	1976	25	F	VF, Cardiac Arrest	Angiography	Operation
Lerberg et al. (16)	1978	10	M	Fatigue	Angiography	Operation
Lerberg et al. (16)	1978	2	M	Death/Myocardial infarction	Autopsy	—
Lerberg et al. (16)	1978	72	M	CHF	Autopsy	—
Bortolotti et al. (17)	1978	22	F	Fatigue, Palpitations	Angiography	Operation
Salomon et al. (18)	1981	35	F	Dyspnea	Angiography	Ligation
Ganz et al. (19)	1981	5	M	None	Angiography	Reimplantation
Chantepie et al. (20)	1982	18	M	None	Angiography	Operation
Mintz et al. (21)	1983	47	M	Angina	Angiography	Operation
van Meurs et al. (22)	1984	13	M	None	Angiography	Operation
Worsham et al. (23)	1985	6	F	None	Echocardiography, Angiography	—
Suzuki et al. (24)	1985	5	M	None	Echocardiography, Angiography	Medical
Saenz et al. (25)	1986	65	F	Myocardial Infarction	Echocardiography, Angiography	Operation
Vairo et al. (26)	1992	.17	M	CHF	Echocardiography, Angiography	Reimplantation
Vogt et al. (27)	1994	1.25	M	—	—	Reimplantation
Dahlstrom et al. (28)	1994	59	F	None	Angiography	Medical
Barth and Apitz (29)	1995	4	F	None	Angiography	Operation
Kautzner et al. (30)	1996	36	F	Angina	Angiography	Operation
Radke et al. (31)	1998	60	F	CHF	Angiography	Reimplantation
Di Luozzo et al. (32)	1998	41	F	Palpitation/angina	—	Reimplantation
Ueeda et al. (33)	1988	48	F	None	Echocardiography, Angiography	—
Vijitbenjaronk et al. (34)	2002	63	M	Angina	Echocardiography, Angiography	Medical
Veselka et al. (35)	2003	36	M	Angina	Echocardiography, Angiography	Reimplantation

AF = atrial fibrillation, CHF = congestive heart failure, F = female, M = male, VF = ventricular fibrillation.

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