*Coronary Anomalies*

# Pitfalls in Diagnosis:

## Suspected Anomalous Origin of the Right Coronary Artery from the Pulmonary Artery

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*Anomalous coronary arteries are rare in the general population. We report the case of a term neonate who underwent an echocardiogram to evaluate a possible patent ductus arteriosus. Unexpectedly, an apparent anomalous origin of the right coronary artery from the main pulmonary artery was detected by surface 2-dimensional transthoracic echocardiography and color-flow Doppler imaging. Because ventricular size and function were normal, the patient ultimately underwent cardiac catheterization to verify the anatomy before proposed surgery. Angiograms showed that the right coronary artery arose from the left anterolateral portion of the mid-ascending aorta. The patient did not require surgery. This case report illustrates pitfalls that can occur in the diagnosis of coronary artery anomalies.*
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Coronary arteries that arise from the pulmonary artery (PA) comprise less than 25% of congenital coronary artery anomalies. Anomalous origin of the left coronary artery from the PA is 10 times more common than origin of the right coronary artery (RCA) from the PA.[1] Anomalous origin of the RCA from the PA has been associated with ischemia, syncope, various cardiomyopathies, and sudden death.[2,3] When the condition is discovered, the only treatment is surgical.

Correct identification of coronary artery origins remains a challenge. Transthoracic echocardiography (TTE) is the primary screening tool for coronary evaluation in children. We report the case of a term newborn who presented with a murmur and subsequently underwent 3 independent echocardiographic examinations, which raised suspicion of an anomalous RCA, quite possibly arising from the main PA. Ultimately, it was determined that the RCA arose anomalously from a point high on the left side of the anterior ascending aorta, thus eliminating the need for surgical intervention.

## Case Report

A 13-day-old boy was referred to our institution for presurgical evaluation. The patient was born at term by normal spontaneous vaginal delivery with a birth weight of 7 lb 14 oz. On the 2nd day of life, a murmur was noted by the primary care physician and TTE was performed for further evaluation. That study revealed a patent ductus arteriosus with a moderate left-to-right shunt, a patent foramen ovale with a left-to-right shunt, and the possibility of an anomalous origin of the RCA from the PA. Otherwise, the echocardiogram showed normal anatomy, including normal ventricular size and systolic function.

The next day, TTE was repeated in order to clarify the coronary origins. This study confirmed that the ductus arteriosus was still patent and again showed 2-dimensional (2D) and color-flow Doppler evidence of an anomalous origin of the RCA from the PA. Of note, a 12-lead electrocardiogram performed that day was normal. Due to these confusing findings, the patient was referred to our institution for further evaluation and management.

The feeding pattern was normal for the age. The patient's mother denied observing any tachypnea, diaphoresis, or cyanosis. The family history was negative for congenital heart disease, genetic disorders, or sudden unexplained deaths.

Auscultation yielded normal results; no murmur was noted. A chest radiograph showed a normal cardiac silhouette with clear, well-expanded lung fields and typical pulmonary vasculature. A 2D TTE in the parasternal short-axis view, however, revealed an anomalous origin of the RCA from the main PA. The right coronary ostium was described as more superior than normal, arising from the anterior surface of the

PA. The color Doppler flow was described as antegrade (from the main PA toward the distal coronary bed) (Fig. 1). Parasternal long-axis images suggested an anomalous course of the RCA with a "ring sign" (Fig. 2). However, the imaging was unclear because the RCA also appeared to have a communication with the ascending aorta and normal antegrade coronary blood flow (Fig. 3). No evidence suggested significant collateral flow within the coronary circulation. Biventricular systolic function was normal, without regional wall-motion abnormalities. The previously noted patent ductus arteriosus was no longer present.

Because the TTE had revealed antegrade diastolic blood flow in the RCA at approximately 2 weeks of age,

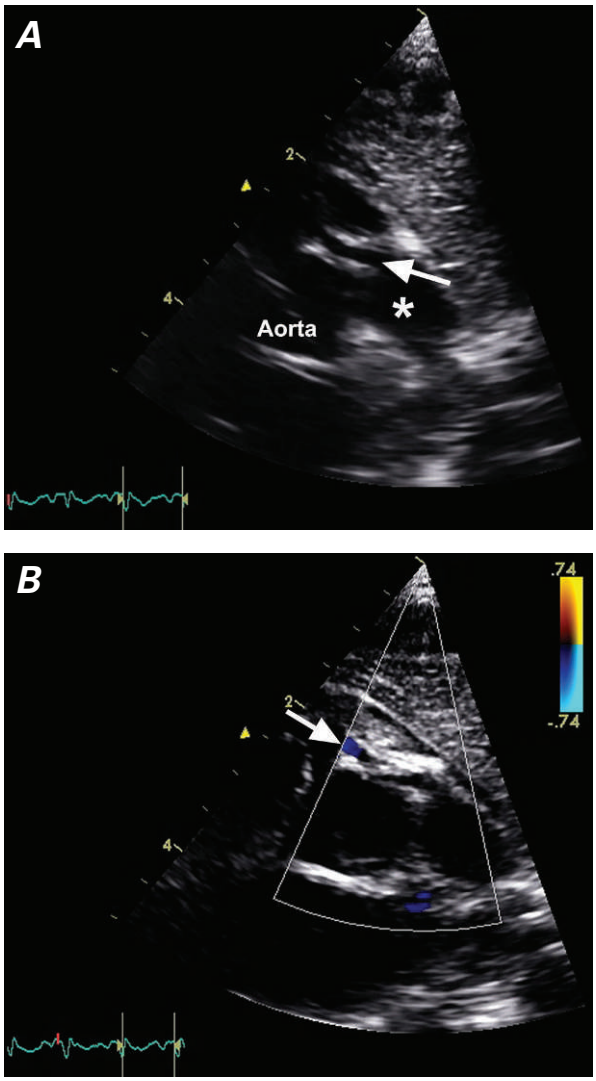


***Fig. 1*** *Two-dimensional transthoracic echocardiograms in **A**) parasternal short-axis view and **B**) color-flow Doppler mode show the right coronary artery ostium (arrow), which appears to originate from the main pulmonary artery (*). It arises slightly more superiorly than normal, from the anterior surface of the pulmonary artery. The color Doppler flow is antegrade from the main pulmonary artery, toward the distal coronary bed.*

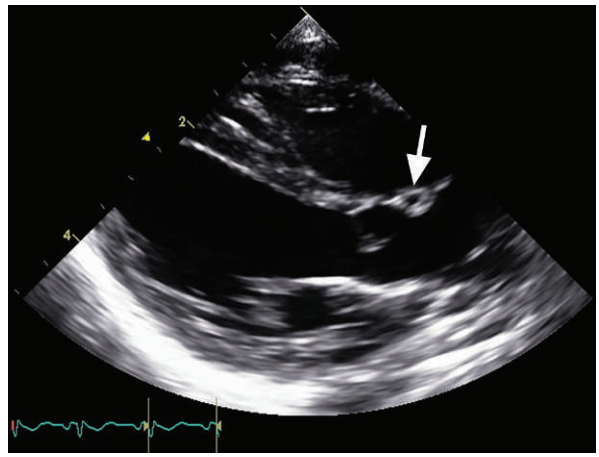

***Fig. 2*** *Two-dimensional transthoracic echocardiogram (parasternal long-axis view) shows a "ring sign" (arrow) just above the sinotubular junction, which suggests an anomalous right coronary origin. Normal left ventricular size is evident.*

and because of the conflicting images of the RCA's origin, cardiac catheterization was conducted before consideration of any surgical intervention. An aortic root angiogram revealed normal left coronary artery origin, size, and distribution (Fig. 4). The RCA was indeed anomalous in origin, but it arose approximately 8 mm above the sinotubular junction on the left anterolateral aspect of the mid-ascending aorta. The vessel coursed anteriorly across the aorta to the right side of the heart. Its size and distribution were otherwise normal, without evidence of stenosis. The ductus arteriosus was absent. The patient was discharged from the hospital the same day with scheduled outpatient follow-up.

## Discussion

The incidence of coronary artery anomalies ranges from 0.6% to 1.3%.[4] Anomalous right origin of the coronary artery from the aorta occurs in only 0.15% of all patients who undergo coronary arteriography. Commonly reported sites for anomalous origin of the RCA include the left sinus of Valsalva, the left main or left anterior descending coronary artery, the PA, and the ascending aorta.[4,5] Anomalous RCAs have been associated with an increased risk of acute myocardial infarction, syncope, and sudden death, particularly if the ostium has an acute takeoff angle from the aorta, or if the vessel courses between the great arteries.[6] Anomalous RCAs have also been associated with other congenital heart defects.[7,8]

In recent decades, coronary anomalies in children have most often been diagnosed with echocardiography. In one large prospective study of children, more than 85% of all coronary anomalies were successfully diagnosed with echocardiography.[9] Although transthoracic echocardiography has the advantage of providing images without the need of general anesthesia, further

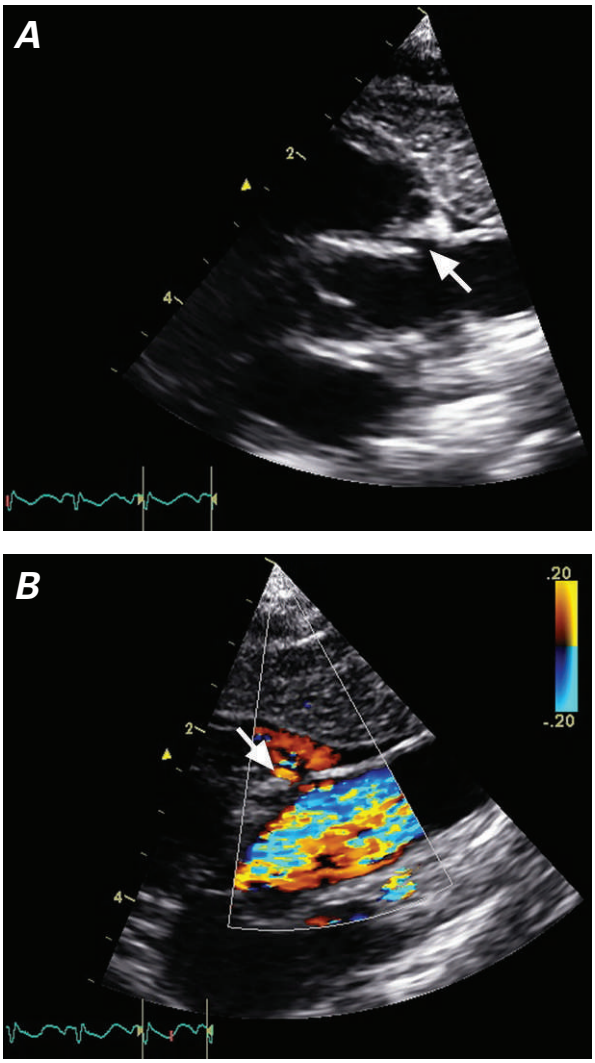


***Fig. 3*** *Two-dimensional transthoracic echocardiogram in **A**) high parasternal long-axis view and **B**) color-flow Doppler mode shows that the right coronary ostium (arrow) arises from the left lateral ascending aorta above the sinotubular junction. The color flow is antegrade from the ascending aorta, toward the distal coronary bed.*

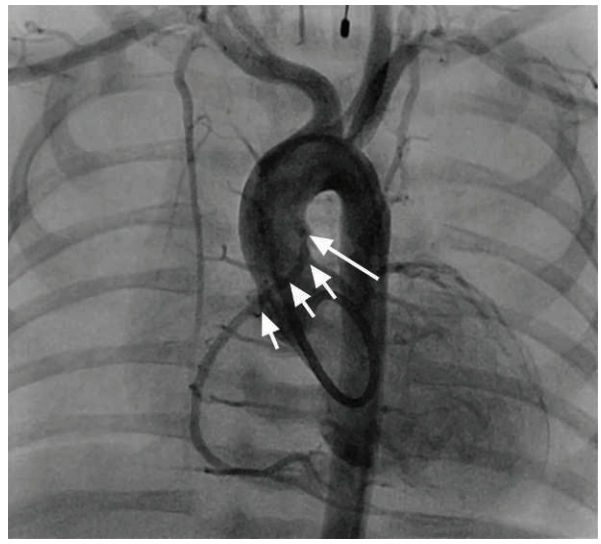

***Fig. 4*** *Aortic root angiogram (anterior/posterior projection) shows that the right coronary artery is anomalous in origin, arising approximately 8 mm above the sinotubular junction on the left anterior aspect of the mid-ascending aorta (long arrow). The vessel courses anteriorly across the aorta to the right side of the heart (short arrows). Its size and distribution are otherwise normal, without evidence of stenosis. Note that the left coronary artery is normal in origin, size, and distribution and that the ductus arteriosus is absent.*

investigation is warranted if the cardiologist is unable to map the coronary arteries satisfactorily by that single means. Multidetector coronary computed tomography remains the gold standard for diagnosing coronary artery anomalies.

At times of increased pulmonary pressure (including the neonatal period) and consequent to pulmonary hypertension of any origin, the coronary blood flow of an anomalous RCA arising from the PA can be antegrade. In our 13-day-old patient, color-flow Doppler images indicated antegrade diastolic flow into the RCA, which suggested persistent elevation of PA pressures. However, other 2D and spectral Doppler echocardiographic evidence (normal right ventricular size, shape, and function; normal estimated right ventricular systolic pressure; and a closed ductus arteriosus) suggested that the pulmonary resistance had already normalized. In patients such as this infant, a 2nd imaging technique (coronary angiography, multidetector coronary computed tomography, or cardiac magnetic resonance) should be considered before surgical intervention.

Due to lower resistance in the pulmonary circulation, coronary anomalies from the PA are associated with retrograde coronary blood flow (into the PA), whereas those from the aorta display antegrade flow (away from the aorta). In the former instance, retrograde blood flow probably will be continuous and can then lead to a coronary steal phenomenon. In the case of the otherwise clinically stable infant described herein, cardiac catheterization provided the best means by which to examine PA pressures (particularly during the diastolic phase), intramural course, steal phenomenon, left- versus right-dominant coronary circulation, and definitive origin.

Historically, anomalous RCA origins above the sinotubular junction have been diagnosed by coronary angiography or by 64-slice multidetector computed tomographic angiography, in preparation for an unrelated surgical procedure.[10] More recently, Thankavel and colleagues[11] have suggested that parasternal long- and short-axis 2D and color-flow Doppler echocardiographic views are not adequate to diagnose anomalous coronary artery origins above the sinotubular junction. They proposed a high parasternal long-axis sweep to increase the diagnostic sensitivity for these anomalies.

In our patient, a high parasternal long-axis sweep was completed. This led to conflicting echocardiographic evidence of the coronary origins. This patient displayed a ring sign in the traditional parasternal long-axis view, as well as 2D echocardiographic evidence of the RCA's origin from the main PA in the traditional parasternal short-axis view. In contrast, the high parasternal long-axis sweep displayed the RCA's origin from the high anterior aorta with normal antegrade blood flow. Therefore, an additional imaging method was indicated to avoid misdiagnosis.

In conclusion, anomalous coronary arteries are rare and clinically significant. Care should be taken to thoroughly evaluate the coronary artery origins from all echocardiographic windows. If origins are unclear, a 2nd imaging technique should be performed before contemplating surgical intervention.

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