



A New Double Coronary Artery Anomaly: The Right Coronary Artery Originating Above the Coronary Sinus Giving off the Circumflex Artery

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This is the first reported case of a coronary artery anomaly in which the right coronary artery originates above the left sinus of Valsalva and also gives off the circumflex system. Use of the aortogram and careful inspection of

the ventriculogram in the right anterior oblique view were most helpful in recognizing the anomaly. The potential pathologic significance of the anomaly and pitfalls of misdiagnosis are discussed.

As the number of coronary arteriograms performed has increased over the past few years, so have the number of coronary artery anomalies described. These anomalies are not of academic interest alone. They have been implicated in a process of accelerated atherosclerosis (1) and syndromes of angina and sudden death in patients with otherwise normal coronary arteries (2). Knowledge of the anatomy and approach to diagnosis can shorten the time required for angiographic examination, reduce the likelihood of restudy (3) and prevent mistakes in the operating room (4).

In an autopsy study of 18,950 patients, Alexander and Griffith (5) identified 54 anomalies and classified them as those of ostial origin (that is, in the contralateral sinus or above the unilateral sinus of Valsalva) and those of distribution (that is, circumflex branch of the right coronary artery), without finding an overlapping case. We describe a case in which the right coronary artery arises above the left sinus of Valsalva and gives off the circumflex branch, combining both classes of anomalies.

Case Report

A 50 year old white man presented with dyspnea on exertion and left pectoral pain at rest and on exertion. His risk factors included obesity and a strong family history of coronary artery disease. A treadmill stress test to less than

90% age-predicted maximal heart rate revealed no ST segment changes or chest pain. He was treated symptomatically without relief of his chest pain, and coronary angiography was performed.

Coronary angiography. The patient underwent catheterization via the right femoral artery. After ventriculography, injections using an 8F, 4 cm, Judkins left coronary catheter revealed a normal left anterior descending artery, but no evidence of the left circumflex artery. A Judkins right coronary catheter was used to explore the right coronary cusp, but no right coronary artery was identified. An aortogram was performed with an 8F pigtail catheter in the left anterior oblique position, with special attention to the ascending aorta. Three sinuses of Valsalva were noted and the aortic valve was normal. The right coronary artery was noted to arise from the ascending aorta approximately 27 mm above the left sinus and anteromedial to the pulmonary artery. From it originated the circumflex artery, which entered the left atrioventricular groove to supply the normal circumflex distribution. Multiple views of this anomalous system were made with a 7F left coronary bypass catheter (Fig. 1). Review of the ventriculogram confirmed this finding. No coronary lesions were found. Subsequently, a maximal thallium stress test was normal.

Discussion

Origin of the circumflex artery as a branch of the right coronary artery accounts for 40 to 54% of coronary artery anomalies found in two large angiographic studies (6,7). Aberrant origin of the right coronary artery from above the contralateral sinus of Valsalva has been described only once (8). The combination is unique to our case.

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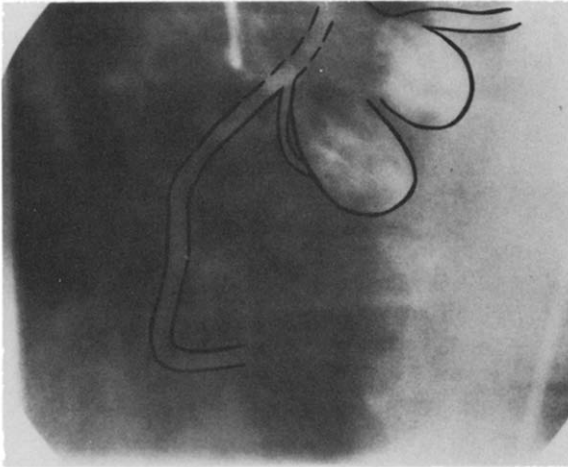


Figure 1. Selective injection of the anomalous right coronary artery in the right anterior oblique position, revealing the course of the left circumflex artery posterior to the aortic root. The sinuses of Valsalva are outlined.

We found the aortogram to be helpful in diagnosing the coronary artery anomaly described. A review of the right anterior oblique ventriculogram also revealed the origin of the right coronary artery to be outside the sinus of Valsalva. Others also found these views to be useful in diagnosing coronary anomalies. Page et al. (6) described the aortic root sign wherein the anomalous left circumflex artery arises from the normal right coronary artery and courses posteriorly behind the aorta, demonstrating a small stub at the root of the aorta on the right anterior oblique ventriculogram. King et al. (8) successfully used aortography to identify an ectopic right coronary artery arising above the left sinus of Valsalva.

Clinical significance. Until recently, the clinical significance of such abnormalities has been thought to be minimal. However, compression of the right coronary artery between the aorta and pulmonary artery, when it arises in the left sinus of Valsalva, has been implicated in at least three cases of sudden death (2). If our patient's right coronary artery arose from within the left sinus of Valsalva, compression between a tense, dilated pulmonary artery and aorta during exercise might be considered a left main coronary equivalent with an increased risk of morbidity and mortality, requiring consideration of coronary artery re-routing. When the right coronary artery arises above the left sinus, as in our patient, dynamic obstruction is less likely to occur (Fig. 2). The pulmonary artery, which lies anterior to the ascending aorta at the level of the sinus of Valsalva (Fig. 3A), directs itself posteriorly as it branches into the left and right pulmonary arteries. A coronary artery that arises above the left sinus (Fig. 3B), therefore, is unlikely to be compressed between a dilated pulmonary artery and aorta during exercise.

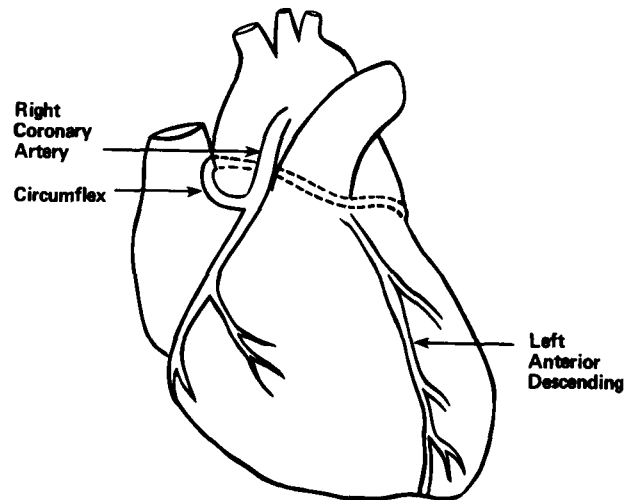
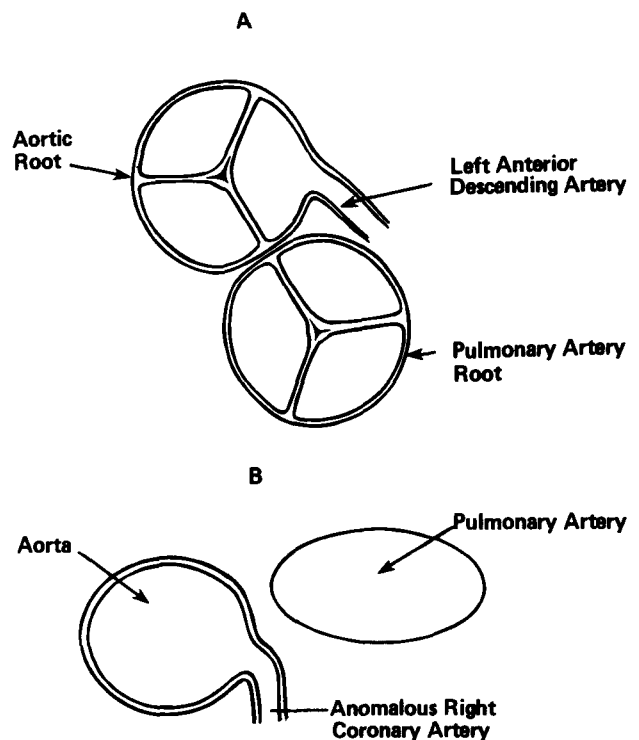


Figure 2. Illustration of the origin and course of the anomalous right coronary artery and the normal left anterior descending artery.

Silverman et al. (1) noted severe proximal atherosclerotic narrowing in the anomalous left circumflex artery of three patients identified from an autopsy study and suggested a

Figure 3. A, Schematic representation of the relation between the aortic root and the pulmonary artery at the level of the origin of the left coronary artery. B, Schematic representation of the relation between the ascending aorta and pulmonary artery at the level of the right coronary artery.



potential for accelerated atherosclerosis in these anomalous vessels. This finding has not been confirmed in our patient or the experience of others (3,6,8).

The importance of identifying an anomalous high take-off of the right coronary artery has been emphasized by Blake et al. (4). They observed that aortotomy or surgical dissection of the aortic root could divide the artery and cross-clamping the aorta may damage the coronary artery. Furthermore, aortic ectasia seen with aging could obstruct the ectopic ostium.

Donaldson and Raphael (3) noted that 38% of their patients with anatomic variants required reinvestigation (four procedures in the case of one patient) before a diagnosis could be made. This delay in diagnosis and treatment causes increased exposure to risk and emphasizes the need for the anticipation of these anomalies and the proper selection of diagnostic techniques to elucidate them.

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