

LETTERS TO THE EDITOR

The Ultimate Cause of Ischemia in a Complex Anomaly

I read with great interest the paper by Eshelbrenner et al. (1) about the exceptional occurrence of 2 coronary anomalies in the same patient, jointly with a coronary complication of mitral valve surgery. The case report's exquisite images are unique and prompt a potentially important point the authors did not make. Although the congenital anomalous origin of the circumflex (Cx) from the right coronary artery (RCA) with a retroaortic course did not by itself have any clinical effect before surgery, accidental Cx ligation occurred during mitral valve annuloplasty. Figure 2 of the Eshelbrenner et al. paper (1) illustrates quite vividly the excellent collaterals that developed over the post-operative years (end-to-end anastomosis from the RCA to the Cx). De novo angina and a positive nuclear stress test at the posterolateral territory in this context must have been mainly caused by the proximal ectopic RCA. Such a vessel indeed originated from the left sinus of Valsalva, probably with a stenotic proximal trunk, that coursed inside the aortic wall, and not "between aorta and pulmonary artery" as is frequently stated (this proximal stenosis apparently was also suggested by angiography). As recently shown by intravascular ultrasound imaging, all cases of ectopic RCA have some degree of stenosis by lateral compression and hypoplasia (2). Such stenosis was the ultimate cause of limited blood flow in the present case because the RCA became the supplier of both the RCA and the Cx territories. This caused the equivalent of significant left main trunk stenosis.

In the post-operative state, after the brilliant initial results of Cx stent angioplasty, the proximal RCA could be a residual or recurrent problem. Specifically, one should be aware that, especially in case of restenosis (likely at an increased risk in this case because a bare-metal stent was used to treat a Cx lesion caused by surgical stitch ligation), symptoms equivalent to those of left main stenosis could recur.

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Reply

We thank Dr. Angelini for his interest in our paper (1) and agree that the ectopic origin and the intramural course of the right coronary artery (RCA) did in fact contribute partly to the clinical presentation of longstanding angina and large inferolateral ischemia. Further evaluation after the stenting of circumflex artery included a significant fractional flow reserve (FFR) of 0.78 in the RCA. We proceeded with primary stenting of proximal RCA with a 4.0 × 15-mm drug-eluting stent (Promus, Boston Scientific, Natick, Massachusetts) followed by post-dilation with a noncompliant balloon and a post-procedure FFR of 0.93. Due to space limitations, this was not included in the initial case report.

The clinical and prognostic implications of an RCA arising from an anomalous cusp and traversing an intramural/inter-arterial course ranges from benign through causing angina to sudden cardiac death (2). To make the distinction and facilitate management, adjunctive modalities such as intravascular ultrasound (3) and FFR (4,5) have been used in a small number of cases. In our patient both involved arteries supplied a large area of myocardium, and thus any significant stenoses would most likely cause ischemia.

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