Long-Term Positive Remodeling of the Right Coronary Artery after Reimplantation from the Pulmonary Artery to the Ascending Aorta

**Miloslav Spacek, M.D.,¹ Theodor Adla, M.D.,²
and Josef Veselka, M.D., Ph.D., F.E.S.C., F.S.C.A.I., F.I.C.A.¹**

ABSTRACT

A 53-year-old woman, nonsmoking patient, with a history of surgically corrected anomalous origin of the right coronary artery from the pulmonary artery 17 years prior to admission, presented to our department complaining of mild, left-sided exertional chest pain for the past 3 months. She underwent a computed tomography examination of the heart and coronary angiography revealing postsurgical changes to the coronary vasculature and severe stenosis of the left circumflex artery, which was successfully treated by percutaneous stent implantation.

KEYWORDS: ARCAPA, steal syndrome, reimplantation, stent

CASE REPORT

A 53-year-old woman, nonsmoking patient presented to our department complaining of mild, left-sided exertional chest pain for the past 3 months. The patient's medical history revealed that, at age 36, she was evaluated for exertional chest pain and diagnosed with anomalous origin of the right coronary artery from the pulmonary artery (ARCAPA) by echocardiography (ECG; Fig. 1) and coronary angiography (Fig. 2). Subsequently, surgical correction was achieved by reimplantation of the dilated right coronary artery (RCA) into the anterior wall of the ascending aorta. Two months post-surgery, a repeated coronary angiography revealed persistent dilatation of the RCA with remarkably slow flow. The left coronary artery (LCA) was normal in appearance. The patient was evaluated annually in follow-up examinations for the past 17 years without any reported complications.

On admission, ECG and laboratory studies were unremarkable. Blood pressure, serum glucose and cholesterol levels were within normal limits, and the patient was not on any medications. The only notable finding revealed during clinical examination was a mild systolic precordial murmur. While the patient was clinically stable, a computed tomography (CT) scan of the heart was performed. The CT scan revealed a tortuous, dilated RCA (Fig. 3), 9 mm in diameter at the origin and 20 mm in diameter at its widest part, which was filled with thrombus (Fig. 4, cross-section level shown in Fig. 5) that gradually narrowed the lumen from 9 mm diameter at its origin to 5 mm in the middle and 4 mm at the distal (inferior) end (Fig. 5). The LCA was suspected to be significantly stenosed in the middle segment of the left circumflex artery (LCx). Subsequent coronary angiography detected the thrombus as a slight defect in the contrast filling along the proximal RCA (Fig. 6), coupled

¹Department of Cardiology, 2nd Medical School, Charles University, Motol University Hospital, Prague, Czech Republic; ²Department of Imaging Techniques, Motol University Hospital, Prague, Czech Republic.

Address for correspondence and reprint requests: Josef Veselka, M.D., Ph.D., F.E.S.C., F.S.C.A.I., F.I.C.A., Department of Cardiology, Cardiovascular Center, Motol University Hospital, Vúvalu 84,

150 06, Prague 5, Czech Republic (e-mail: veselka.josef@seznam.cz).

Int J Angiol 2011;20:117–120. Copyright © 2011 by Thieme Medical Publishers, Inc., 333 Seventh Avenue, New York, NY 10001, USA. Tel: +1(212) 584-4662.

Published online: June 3, 2011.

DOI: <http://dx.doi.org/10.1055/s-0031-1279684>.

ISSN 1061-1711.

Figure 1 Presurgical transesophageal echocardiography, showing the right coronary artery (RCA) originating from the pulmonary artery (AP) and dilated left main coronary artery (LMCA) originating from the aorta, left atrium (LA).

with no signs of slow flow. An angiogram of the otherwise normal LCA confirmed severe stenosis of the middle segment of the LCx, which was treated with percutaneous stent implantation. Subsequently, all symptoms disappeared. The patient was discharged on aspirin, clopidogrel, and rosuvastatin.

Figure 2 Presurgical coronary angiography of the LCA, with collateral retrograde filling of the RCA and the pulmonary artery. LCA, left coronary artery; RCA, right coronary artery.

Figure 3 CT scan (volume rendering technique) of the heart with the RCA reimplanted into the aorta. CT, computed tomography; RCA, right coronary artery.

DISCUSSION

Isolated ARCAPA is a rare congenital coronary anomaly. Although the incidence rate of ARCAPA is estimated to be ~0.002%, many patients are asymptomatic, and the true prevalence of ARCAPA is likely underestimated. The pathophysiological effects of ARCAPA are related to the direction of blood flow in the coronary artery. In fetuses and newborns, blood flows from the pulmonary artery into the anomalous coronary artery because of high pulmonary vascular resistance. During the first few days of life, pulmonary vascular resistance falls, resulting in

Figure 4 CT scan of reconstructed cross-section of the RCA at its widest portion. CT, computed tomography; RCA, right coronary artery.

Figure 5 CT angiography (curved multiplanar reconstruction) of the RCA. CT: computed tomography; RCA, right coronary artery.

three possible scenarios: (1) insufficient collateralization between the RCA and the LCA will result in ischemia and death; (2) adequate collateralization may also result in ischemia because of a steal phenomenon; or (3) massive collateralization may enable maintenance of adequate myocardial perfusion even in the face of a steal phenomenon. Usually, individuals with right dominant coronary circulation do not tolerate ARCAPA.¹

Because adverse outcomes (including increased risk for myocardial infarction and sudden cardiac death without any preceding symptoms) have been described repeatedly in ARCAPA patients, surgical correction is recommended whenever this anomaly is recognized. Three different surgical procedures have been previously advocated: RCA ligation at its origin, with or without a venous aorto-coronary graft to its distal end; or direct reimplantation of the RCA into the anterior wall of ascending aorta. However, the long-term patency of direct reimplantation is thought to be better than that of a venous bypass and therefore has more proponents.²

Because reports describing ARCAPA follow-up are rare, there is only limited knowledge about the long-term changes to coronary vasculature after surgical correction. Nevertheless, surgical and pathology reports describe the anomalous RCA to be thin-walled, dilated, and vein-like in character; therefore, unlike the LCA, it is thought that the RCA does not tend to normalize in diameter after surgical correction of ARCAPA.^{1,3}

Figure 6 Coronary angiography of the RCA, showing gradual narrowing and slight thrombotic defects (arrows). RCA, right coronary artery.

CONCLUSION

This case report illustrates the long-term changes to coronary vasculature after surgical correction of ARCAPA; the LCA tends to normalize its diameter and flow, whereas the RCA tends to remain dilated for the long term and undergoes positive remodeling.

NOTE

The authors disclaim any financial interests with respect to publication of this article.

REFERENCES

1. Williams IA, Gersony WM, Hellenbrand WE. Anomalous right coronary artery arising from the pulmonary artery: a report of 7 cases and a review of the literature. *Am Heart J* 2006; 152:1004, e9–e17
2. Veselka J, Widimský P, Kautzner J. Reimplantation of anomalous right coronary artery arising from the pulmonary trunk leading to normal coronary flow reserve late after surgery. *Ann Thorac Surg* 2003;76:1287–1289
3. Su JT, Krishnamurthy R, Chung T, Vick GW III, Kovalchin JP. Anomalous right coronary artery from the pulmonary artery: noninvasive diagnosis and serial evaluation. *J Cardiovasc Magn Reson* 2007;9:57–61