Single Coronary Artery With Subsequent Coursing of Right Coronary Artery Between Aorta and Pulmonary Artery

Fractional flow reserve of the anomalous artery guiding the treatment

Nurten SAYAR,1 MD, Sait TERZI,1 MD, Tamer AKBULUT,1 MD, Tuba BILSEL,1 MD, Mehmet ERGELEN,1 MD, Lutfu ORHAN,1 MD, Nazmiye CAKMAK,1 MD, and Kemal YESILCIMEN,1 MD

SUMMARY

A single coronary artery with an anomalous origin between the pulmonary artery and aorta is an unusual congenital anomaly. We present a 60 year old female patient with stable angina pectoris. Her coronary angiogram revealed that the right coronary artery originated from the left main stem and coursed between the great vessels. There was 90% stenosis in the left anterior descending coronary artery. We performed a fractional flow reserve study of both the anomalous origin artery and stenosed vessel. Fractional flow reserve study of the anomalous RCA did not reveal functional ischemia. We did not refer the patient to coronary artery bypass grafting but instead performed percutaneous coronary revascularization of the LAD. The patient is alive and has been free of symptoms during the 1 year follow-up.  (Int Heart J 2005; 46: 317-322)

Key words: Coronary anomaly, Single coronary artery, Fractional flow reserve

A single coronary artery is an unusual congenital anomaly of the coronary arteries where only one coronary artery arises from the aortic trunk by a single coronary ostium, supplying the entire heart. As an isolated finding it occurs in approximately 0.024% to 0.044% of the population. In association with certain other congenital anomalies, however, it is found more frequently. According to Banchi, the first case of a single coronary artery was reported by Thebesius in 1716. Until 1950 no more than 45 cases had been published, all discovered at autopsy.

In 1979 Lipton, et al proposed a very useful angiographic classification, further modified in 1990 by Yamanaka and Hobbs. In this classification, a first division is made between the 'R' right-type and the 'L' left-type according to the
site of origin of the single coronary artery, ie, in the right or left sinus of Valsalva. Every case is then designated as belonging to group I, II, or III depending on the anatomical course of the artery.

We present a patient in whom the RCA arises from the left main stem and traverses between the aorta and pulmonary artery, designated as L II B according to the Lipton classification. There is 90% stenosis in the LAD. This is a rare case in the literature where FFR was used to guide the therapeutic modality.

CASE

A 60 year old female patient with a history of hyperlipidemia and hypertension was referred for preoperative cardiac consultation before a laparoscopic cholecystectomy operation. She had stable angina pectoris Class 2. She was on acetylsalicylic acid, nitrate, beta blocker, and ACE inhibitor therapy. She underwent exercise technetium myocardial perfusion scintigraphy (MPS) which was positive for severe anterior and anteroseptal ischemia. Coronary angiography was subsequently performed which revealed a single coronary artery where there is anomalous origin of the RCA that originated from the left main stem (Figure 1A). There was also 90% discrete stenoses in the mid LAD. Left ventriculography and echocardiography showed normal left ventricular function with no wall motion abnormality. We performed electron beam computed tomography (EBT) to determine the course of the RCA. EBT revealed that the RCA coursed between the aorta and right ventricular outflow tract (RVOT) (Figure 1B). There was no apparent extraluminal compression of the RCA between the great vessels. EBT failed to show occlusion in the proximal LAD, suggesting only that there was

Figure 1. A: Coronary angiography of the patient. RCA originates from the left main stem. There is 90% stenosis in the mid LAD. B: EBT of the patient showing the course of RCA. RCA courses between the aorta and right ventricular outflow tract.

EBT = electron beam computed tomography; RVOT = right ventricular outflow tract; RCA = right coronary artery; LAD = left anterior descending artery; IM = intermedier artery; CX = circumflex artery).
apparent mural irregularities in the proximal and mid LAD. According to MPS, there was significant anterior and anteroseptal ischemia but no exercise-induced ischemia in the territory of the RCA. We tested if the MPS result showing no ischemia in the RCA territory could be reproduced using fractional flow reserve (FFR). If the FFR of RCA is hemodynamically significant, the patient would be referred for surgery. Before the procedure, the purpose of the study and all stages of the procedure were explained to the patient and informed consent was obtained. The FFR of the RCA was determined at basal heart rate, after adenosine infusion, and during atrial pacing. A pressure transducer was placed in the distal RCA, distal to the point where it courses between the great vessels. The basal FFR of the RCA in a resting state with a HR of 85 beat/min was 0.95 (Figure 2A), and 0.90 after direct intracoronary infusion of 30 µg adenosine (Figure 2B). The FFR was 0.92 after pacemaker-induced tachycardia for 3 minutes when the HR was 150 beats/min (Figure 2C). The patient did not complain of chest discomfort during tachycardia but there was ST segment depression on V₂ through V₆. Intracardiac right and left pressures before and after tachycardia are summarized in the Table. There was no hemodynamically significant change in RCA flow assessed by FFR in the three conditions. We performed direct stenting of the LAD using a 3 × 15 mm Ephesos stent without any complications. The FFR of the LAD at basal state was 0.56, which was significant (Figure 3A). After stenting, the FFR was 0.94 after adenosine infusion (Figure 3B). We performed an exercise treadmill test at 1 month after percutaneous coronary intervention (PCI). There was neither angina nor ischemic ECG changes at 12 MET workload. The sixth month

**Figure 2.** Fractional flow reserve of RCA at A (basal), B (after adenosine infusion), and C (pace-maker induced tachycardia state). RCA = right coronary artery
MPS study result was also normal and in agreement with the FFR results. The patient is alive and has been free of symptoms throughout the 1 year follow-up period.

**DISCUSSION**

It is well established that origin of the left main coronary artery from the right sinus of Valsalva with subsequent coursing between the aorta and pulmonary trunk is associated with sudden death. However, the association of risk of sudden death and anomalous origin of the RCA from the left sinus of Valsalva is not so clear. The mechanism by which origin of the right coronary artery from the left sinus may cause cardiac dysfunction is unclear. Possibly, the anomalous artery is compressed during its course between the aorta posteriorly and the pulmonary trunk anteriorly in a manner similar to that proposed for the LMCA. A more likely possibility is diminished flow into the anomalous right coronary artery due to its upright slit-like origin from the aorta and its peculiar orientation to the mid portion of the lumen of the aorta. When both the pulmonary trunk and aorta dilate during exercise, the slit-like ostium also prevents blood within the

**Table.** Intracardiac Pressures of the Patient in Basal and Pacemaker-induced Tachycardia States

<table>
<thead>
<tr>
<th>Pressure (mmHg)</th>
<th>Resting</th>
<th>Tachycardia</th>
</tr>
</thead>
<tbody>
<tr>
<td>RA</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>RV (systole/diastole)</td>
<td>20/6</td>
<td>37/5</td>
</tr>
<tr>
<td>PA (systole/diastole)</td>
<td>20/12</td>
<td>37/13</td>
</tr>
<tr>
<td>PCW</td>
<td>12</td>
<td>13</td>
</tr>
</tbody>
</table>

RA = right atrium, RV = right ventricle, PA = pulmonary artery, PCW = pulmonary capillary wedge.

**Figure 3.** Fractional flow reserve of LAD in the basal state (A) and after stent deployment and intracoronary adenosine infusion (B). LAD = left anterior descending artery.
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Fractional flow reserve of coronary anomaly. The aortic lumen from flowing into the right coronary artery during ventricular diastole. The ostium of an anomalous artery, the RCA in this case, is not in the aorta but rather originates from the LMCA. Therefore, the slit-like origin is not relevant in our case. Our concern was the course of the RCA between the great vessels. However, when all cases were reviewed, we were not certain CABG was suitable for this patient. Roberts, et al reported 10 necropsy patients whose RCA originated from the left sinus of Valsalva. The course of the artery between the great vessels was not stated. All patients were less than 50 years of age. Seven of the patients died of noncardiac causes. One 17 year old patient died of sudden death while playing basketball. A 49 year old patient had angina pectoris for 12 months and died during coitus. The third patient was 23 years of age and presented initially with syncope provoked while running and ventricular tachycardia was documented on many occasions. In the series of Kragel et al, 25 cases had anomalous origin of the right coronary artery. The cause of death was a coronary anomaly in 8 patients (32%). The age range was 16 to 49, with an average of 26.5 years. Of these 8 patients, 5 had no signs or symptoms of cardiac disease while alive. All of these patients were young, with the oldest case being 40 years of age. Two patients had angina pectoris and 1 patient had ventricular tachycardia. Compared to the cases of Roberts and Kragel who died of sudden death, our patient was older (60 years of age) and a housewife with a low likelihood of vigorous exercise. She had no history of syncope. She had angina pectoris, the origin of which we believe was the LAD not RCA as confirmed by both MPS and FFR. There is a paucity of experience with FFR in coronary anomalies. Angelini, et al used FFR during adenosine provocation testing in patients with a coronary artery originating from the opposite sinus (ACAOS) and had results within the normal limits (FFR > 0.9). Cheitlin, et al stated that the normal thickening and stiffening of the aortic wall that occurs with aging may account for the relatively benign behavior of ACAOS in older patients. Therefore, we did not find enough evidence in the literature to refer this patient to CABG. However, there are limitations in this case: Firstly, we did not have the IVUS study of the anomaly, and secondly FFR has never been validated for choosing a treatment option.

This case is interesting in several ways. It is a single coronary artery, the anomalous coronary artery courses between the great vessels, FFR was used for the first time to guide the treatment of the anomalous artery, and percutaneous coronary intervention was applied in a single coronary artery case. With this therapeutic option, the patient did not need to undergo major cardiac surgery and has been free of symptoms during the 1 year follow-up.
REFERENCES