Anomalous origin of the right coronary artery: Right internal thoracic artery to right coronary artery bypass is not the answer

Lynn M. Fedoruk, MD, John A. Kern, MD, Benjamin B. Peeler, MD, and Irving L. Kron, MD

Objective: Anomalous origin of the right coronary artery from the opposite sinus of Valsalva can be a lethal congenital anomaly. Right internal thoracic artery grafting to the right coronary artery is prone to fail in this circumstance. We sought to describe alternative surgical techniques.

Methods: Retrospective analysis identified 5 adult and pediatric patients in our database. We reviewed the surgical techniques used to repair this anomaly. On the basis of our experience, we describe our management technique.

Results: There were no operative deaths, and postoperative computed tomographic scans demonstrated widely patent repairs in all patients. Two patients with previous right internal thoracic artery to right coronary artery grafts presented with occlusion of the right internal thoracic artery. Short-term follow-up demonstrated continued patency.

Conclusion: Right internal thoracic artery grafting fails in this circumstance, and alternative surgical options provide a good outcome.

Anomalous aortic origin of a coronary artery from an opposite sinus of Valsalva is a rare and sometimes lethal congenital anomaly. Isolated coronary artery anomalies have been described in up to 1% of all patients undergoing cardiac catheterization. Autopsy series have placed the incidence at approximately 0.3% for all coronary anomalies. Anomalous origin of a coronary artery from the opposite sinus of Valsalva is significantly less common, with the estimated incidence of 0.03% to 0.05% for the left origin arising from the right sinus of Valsalva and an estimated incidence of 0.05% to 0.1% for the right origin arising from the left sinus of Valsalva.

The most concerning issue related to this condition is the association with sudden death, especially in young asymptomatic athletes. In one autopsy series 28.8% (15/52) of patients died as a result of the right coronary artery (RCA) arising from the left sinus, with 13 of the 15 experiencing sudden death. In another study 32% of patients experienced sudden death associated with this anomaly. In addition to sudden death, angina, palpitations, syncope, and dyspnea are associated with this condition.

The choice of treatment for this congenital anomaly is still controversial; however, because of the propensity for sudden death, especially in association with exercise in younger individuals, most of the literature advocates definitive surgical correction. Multiple options have been advanced for correction of this condition, including coronary artery bypass grafting (CABG), reimplantation of the coronary ostia, and unroofing of the coronary artery (excision of the common wall between the aorta and the RCA). This report summarizes our observations and experience of the various techniques, particularly outcomes associated with CABG.
In 4 of the 5 procedures, the RCA was then unroofed along the thin common wall with the aorta with a 15 blade.10,11 The artery was unroofed for a distance of 1.0 to 1.5 cm. The circumference of the tract was then reapproximated (the intima of the RCA to the intima of the aorta) with 6-0 Prolene sutures (Ethicon, Summersville, NJ), creating a neo-ostium farther along the tract. In the first 3 unroofing procedures, a running technique was used for suturing, and in the most recent procedure, sutures were placed in an interrupted fashion. A coronary probe was passed into the neo-ostium to confirm patency. The aorta was then closed in a 2-layer fashion, and after deairing, the patients were weaned from CPB. Unfortunately, in one case, a nonredo operation, there was evidence of right heart ischemia after weaning from CPB that necessitated reinitiating CPB, recrossclamping, and placement of a saphenous vein graft to the distal RCA. No further ischemia was noted.

In the first procedure, on inspection of the anomalous RCA orifice, a decision was made to reimplant the artery as opposed to unroofing it because there was concern that commissure of the left/right aortic valve cusps would be compromised, leading to the potential for aortic insufficiency.10 In this case the RCA was extensively dissected and then divided at its ostium, with the resultant hole in the aorta being oversewn with 5-0 Prolene sutures (Ethicon). Visualizing the interior of the right sinus of Valsalva, a neo-ostium was then created with a 4.8-mm aortic punch in a position that was away from the pulmonary artery. Care was taken to ensure the new course of the RCA was tension free and would not cause kinking. The ostium of the RCA was spatulated to ensure a cobra hood–type proximal anastomosis, and a standard end-to-side anastomosis was then performed with 7-0 Prolene sutures (Ethicon).

Patients were weaned from CPB in the usual fashion. All patients recovered in the thoracic and cardiovascular postoperative unit and were discharged home after short hospital stays with appropriate pain medication and antiplatelet agents. Before discharge, CT angiography was performed to assess patency of the repair.

Results
The study cohort consisted of 5 patients who underwent surgical repair. The average age was 35.0 ± 16.7 years. Preoperative characteristics are listed in Table 1. Two had previously undergone a right internal thoracic artery (RITA) to RCA bypass graft without proximal ligation of the native coronary artery. In both cases, the previous symptoms recurred (average time symptom free, 7 ± 5 months), with repeat cardiac catheterization demonstrating occluded RITAs.

Operative characteristics are listed in Table 2. Average cardiopulmonary bypass time was 82.4 ± 35.8 minutes, and the average crossclamp time was 51.0 ± 18.8 minutes. In each case the anomalous right ostium was located in the left sinus of Valsalva anterior, superior, or both to the left coronary ostium (Figure 1). In 2 cases the orifice was small or slit like.

Postoperative CT angiography was performed in each patient and demonstrated a widely patent neo-ostice removed from the pulmonary artery (Figure 2), including the patient who required saphenous vein bypass. In retrospect, the right ventricular dysfunction observed in this patient was most likely caused by air. There were no operative mortalities. Average length of stay was 5 ± 2 days. No
The patient has demonstrated recurrent symptoms, although admittedly, none have been followed further than 1 year.

**Discussion**

Anomalous coronary ostia are a recognized cause of sudden death, especially associated with high-intensity exercise in young adults. Traditional diagnostic techniques, such as coronary angiography and, to a lesser extent, transesophageal echocardiography, are invasive and ultimately underused. Improvements in noninvasive diagnostic techniques, such as transthoracic echocardiography and CT angiography, have increased the ability to easily and safely screen for the condition, leading to increased rates of diagnosis.

Sudden death is thought to be associated with restriction of flow down the anomalous artery, causing myocardial ischemia and ventricular arrhythmias, especially when the anomalous coronary artery courses between the great vessels (aorta and pulmonary artery). At present, the mechanisms that lead to myocardial ischemia are unclear, but several potential mechanisms have been proposed.

The coronary ostial dimensions might play a role. A slit-like opening by itself might be flow limiting and with exercise might become more restrictive, especially if there is a flap-like opening that could act as a 1-way restrictive valve as aortic pressures increase with exercise. An acute angulation of the opening as the aberrant vessel exits the aorta (as opposed to the normal perpendicular exit of the RCA from its normal position) might lead to alteration of flow patterns, with restrictions increasing during increased aortic pressures associated with exercise. It has also been hypothesized that the course of the anomalous coronary artery between the 2 great vessels might lead to compression; however, it is more likely that increased aortic wall tension relative to the wall tension in the coronary artery results in coronary deformation and decrease in coronary flow, especially at times of increased aortic pressure (ie, aerobic exercise). It has been demonstrated by using intravascular ultrasonography that there is a 30% to 50% reduction in the luminal area of the coronary artery in systole.

Ultimately, it is important to understand the pathophysiology to attempt an effective treatment of this condition. Multiple therapeutic options have been suggested. Medical therapy with β-blockade has been described with some efficacy in case reports. This decreases aortic wall tension (decreased dP/dT) and myocardial oxygen demand and palliates myocardial oxygen reserve issues; however, it does not treat the cause. Percutaneous stenting of the anomalous coronary artery as it courses between the great vessels has also been advocated by some physicians, but this cannot effectively treat ostial issues that might be present and leaves a long stent length that might be problematic in the future because it might be particularly prone to occlusion.

---

**TABLE 1. Preoperative characteristics**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Age (y)</th>
<th>Sex</th>
<th>Symptoms</th>
<th>ECG</th>
<th>Echo</th>
<th>MIBI</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>33</td>
<td>M</td>
<td>Syncope with exertion</td>
<td>Normal</td>
<td>LVH</td>
<td>Normal</td>
</tr>
<tr>
<td>2</td>
<td>51</td>
<td>F</td>
<td>Ventricular fibrillation arrest</td>
<td>Normal after arrest</td>
<td>Hypokinesis of apical septum</td>
<td>Normal</td>
</tr>
<tr>
<td>3</td>
<td>26</td>
<td>F</td>
<td>Angina, dyspnea</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>4</td>
<td>52</td>
<td>F</td>
<td>Angina</td>
<td>Normal</td>
<td>Normal</td>
<td>Normal</td>
</tr>
<tr>
<td>5</td>
<td>13</td>
<td>M</td>
<td>Angina</td>
<td>LVH</td>
<td>LVH and anomalous RCA</td>
<td>Normal</td>
</tr>
</tbody>
</table>

ECG, Electrocardiogram; Echo, transthoracic echocardiogram; MIBI, technetium sestimibi nuclear study; CTA, computed tomographic angiogram; LVH, left ventricular hypertrophy; NA, not applicable or not performed; RCA, right coronary artery; OPCAB, off-pump coronary artery bypass graft; RITA, right internal thoracic artery; LAD, left anterior descending artery; PCI, percutaneous intervention.

**TABLE 2. Operative characteristics**

<table>
<thead>
<tr>
<th>Patient no.</th>
<th>Anatomic finding</th>
<th>Redo</th>
<th>Previous procedure</th>
<th>CPB time</th>
<th>Crossclamp time</th>
<th>CABG Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Slit ostium immediately anterior to the left os</td>
<td>Yes</td>
<td>RITA to RCA</td>
<td>83</td>
<td>48</td>
<td>No Reimplantation</td>
</tr>
<tr>
<td>2</td>
<td>Right os medial to left os in the same proximal/distal plane</td>
<td>No</td>
<td>LAD stent</td>
<td>45</td>
<td>32</td>
<td>No Unroofing</td>
</tr>
<tr>
<td>3</td>
<td>Right os superior and anterior to left os</td>
<td>Yes</td>
<td>RITA to RCA</td>
<td>97</td>
<td>76</td>
<td>No Unroofing</td>
</tr>
<tr>
<td>4</td>
<td>Right os superior and anterior to left os, small right os</td>
<td>No</td>
<td>Nil</td>
<td>134</td>
<td>64</td>
<td>Yes Unroofing + CAB</td>
</tr>
<tr>
<td>5</td>
<td>Right os superior and anterior to left os</td>
<td>No</td>
<td>Nil</td>
<td>53</td>
<td>35</td>
<td>No Unroofing</td>
</tr>
</tbody>
</table>

Redo, Reoperation; CPB, cardiopulmonary bypass; CABG, coronary artery bypass graft; os, coronary ostium; RITA, right internal thoracic artery; RCA, right coronary artery; LAD, left anterior descending artery.
Multiple surgical therapies have also been advanced. CABG, coronary artery reimplantation, and ostial unroofing have been advocated. As demonstrated in 2 of our patients, CABG, especially without proximal ligation of the native coronary artery, leads to an unsatisfactory result. Other series have also reported graft failure. Although this evidence is anecdotal, serious thought must be given to the potential for graft failure. Because most of the patients that present for surgical correction are young, some advocate that bypass with an internal thoracic artery (ITA) increases the potential for long-term patency, especially relative to placement of a saphenous vein graft. Unfortunately, as shown separately by both Sabik and colleagues and Berger and associates, without proximal ligation, the arterial graft often fails because of competitive flow. Ligation of the native RCA is of significant concern because we and others believe that the initial flow from an ITA graft might not be enough to compensate for acute ligation of a patent vessel, leading to an increased incidence of hypoperfusion syndrome, ischemia, and even mortality. This has been demonstrated when minimally diseased vein grafts are replaced by an ITA.

Reimplantation has been advocated, especially if there is concern related to the position of the coronary ostia relative to the aortic valve commissures. Unroofing in the setting of involvement of the valve commissure might present the hazard of causing aortic insufficiency, creating a larger problem. In addition, it should be noted that some anomalous coronary arteries are not intramural (within the aortic wall) but rather distinct arteries. This can be determined by carefully examining the preoperative imaging. In this instance reimplantation would be the treatment of choice.

In our series our first patient required reimplantation to avoid damage to the aortic valve. The dilemma with advo-
cating reimplantation as a primary repair technique involves concerns over the increased complexity of the procedure. A significant length of the RCA must be dissected to allow for manipulation, increasing the potential for injury. This dissection might also be exceedingly difficult given the intramural course of the vessel that can preclude development of Carrel patch. Neo-ostial obstruction might be precipitated because of the lack of a coronary button for fashioning an anastomosis. Precise placement of the neo-ostia must occur due to decrease risks associated with kinking and flow disruption from a less than perfectly reimplanted vessel.

Coronary artery unroofing, as described, involves creating an enlarged neo-orifice in a position removed from the pulmonary artery. This is often easily performed because of the long, thin common wall shared between the anomalous RCA and the aorta. This addresses the majority of the potential causes discussed above. The ostium is enlarged, the angulation is corrected, and the arterial path is removed from passing between the great vessels, as well as from being intramural within the aortic wall. We believe that if anatomy permits, this is the best approach to correct this anomaly.

Multiple therapies, medical, percutaneous, and surgical, have been advocated for this rare yet potentially lethal condition. CABG of the RCA with an RITA is prone to failure, particularly without proximal ligation, which has significant concerns associated with it. We believe that if anatomically feasible, unroofing of the anomalous RCA provides the best solution for this congenital cardiac abnormality.

References