

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

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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.¹ Autopsy series have placed the incidence at approximately 0.3% for all coronary anomalies.² 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.³

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,⁴ with 13 of the 15 experiencing sudden death. In another study 32% of patients experienced sudden death associated with this anomaly.⁵ In addition to sudden death, angina, palpitations, syncope, and dyspnea⁶ 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),⁷ reimplantation of the coronary ostia,^{8,9} and unroofing of the coronary artery (excision of the common wall between the aorta and the RCA).¹⁰⁻¹² This report summarizes our observations and experience of the various techniques, particularly outcomes associated with CABG.

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Received for publication Aug 4, 2006; revisions received Oct 3, 2006; accepted for publication Oct 9, 2006.

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J Thorac Cardiovasc Surg 2007;133:456-60
0022-5223/\$32.00

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doi:10.1016/j.jtcvs.2006.10.011

Abbreviations and Acronyms

CABG	= coronary artery bypass grafting
CPB	= cardiopulmonary bypass
CT	= computed tomography
ITA	= internal thoracic artery
RCA	= right coronary artery
RITA	= right internal thoracic artery

Materials and Methods

After institutional review board approval was obtained, a review of the University of Virginia cardiac surgical database identified 5 consecutive patients (3 female and 2 male patients) who underwent repair of an anomalous RCA arising from the left sinus of Valsalva between August 2004 and April 2006. A retrospective review of medical records and preoperative diagnostic testing was performed.

All patients had preoperative technetium 99m sestimibi studies performed, and 4 of 5 had computed tomographic (CT) angiography. Four of 5 had cardiac catheterization for delineation of the anatomy. All had preoperative echocardiograms, which were reviewed.

The surgical technique for our cohort is described below. Four patients underwent unroofing (with 1 requiring a subsequent bypass), and 1 had reimplantation. The type of repair was determined preoperatively based on our impressions from the CT scan, echocardiogram, or both. The location of the anomalous coronary artery with respect to the aortic valve commissures guided the type of repair used. If there was potential for aortic valve compromise, then the reimplantation technique was used. If there was no common wall between the 2 vessels, then reimplantation would be used.

The technical aspects of the procedure are as follows. Patients were started on cardiopulmonary bypass (CPB) by using standard cardiac surgery techniques. A left ventricular venting cannula was placed through the right superior pulmonary vein for visualization. If the patient was undergoing a redo operation, as much of the dissection was performed as possible before placing any cannulas. Cannulation then occurred, the patient was started on CPB, and the remainder of the dissection performed.

After crossclamping, the heart was arrested with cold blood antegrade cardioplegia, retrograde cardioplegia, or both. Cold blood cardioplegia was reinforced on average every 20 minutes. A "hot shot" was administered before removal of the crossclamp.

With the aortic crossclamp in place, the aorta was opened in a transverse fashion approximately 2 cm above the sinotubular junction. The aortotomy was then extended parallel to the annulus of the aortic valve. At this point, the coronary ostia were identified. The right coronary ostium was identified, and a right-angle clamp was gently placed to confirm the course of the artery.

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, Summerville, 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 reinstating 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-orifice 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

TABLE 1. Preoperative characteristics

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

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.

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.⁴ 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 echocardiography¹¹ 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.¹³ An acute angulation of the opening as the aberrant vessel exits the aorta (as opposed to the normal perpendic-

ular 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.¹³ 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).⁴ 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.¹⁴

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.¹⁵ 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,⁷ 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.^{16,17}

TABLE 2. Operative characteristics

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

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.

TABLE 1. Continued

CTA	Cardiac catheterization	Prior operation	Symptom-free period	Recurrent symptom	Repeat catheterization
NA	Normal except anomalous RCA	OPCAB RITA to RCA	5 mo	Angina	RITA occlusion
Anomalous RCA	70% LAD occlusion and anomalous RCA	LAD coated stent	NA	Angina after PCI	NA
Anomalous RCA	Normal except anomalous RCA	RITA to RCA	7 mo	Angina	RITA occlusion
Anomalous RCA	Mild CAD and anomalous RCA	None	NA	NA	NA
Anomalous RCA	NA	None	NA	NA	NA

Multiple surgical therapies have also been advanced. CABG,⁷ coronary artery reimplantation,^{8,9} and ostial unroofing¹⁰⁻¹² 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.⁷ 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.¹⁸ Unfortunately, as shown separately by both Sabik and colleagues¹⁹ and Berger and associates,²⁰ 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.²¹ This has been demonstrated when minimally diseased vein grafts are replaced by an ITA.²¹

Reimplantation has been advocated,^{8,9,22} 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.¹¹ 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-



Figure 1. Preoperative computed tomographic angiogram demonstrating an anomalous right coronary artery originating from the left sinus of Valsalva. Note the acute angle of takeoff of the right coronary artery and the course of the artery adjacent to the wall of the aorta and in between the root of the aorta and main pulmonary artery (arrow).

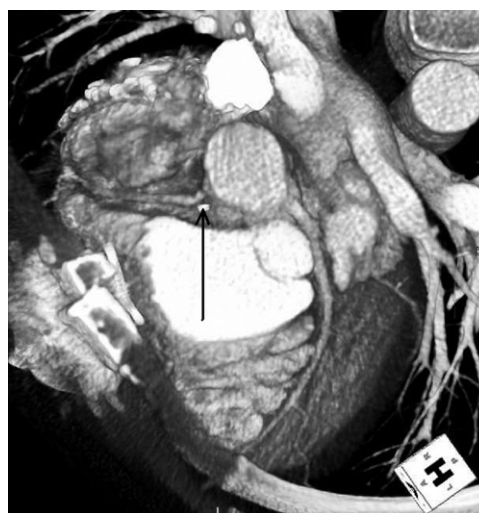


Figure 2. Postoperative computed tomographic angiogram reconstruction. Note the widely patent neo-orifice of the right coronary artery and lack of potential for great vessel compression (arrow).

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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 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

1. Yamanaka O, Hobbs RE. Coronary artery anomalies in 126,595 patients undergoing coronary arteriography. *Catheter Cardiovasc Diagn.* 1990;21:28-40.
2. White NK, Edwards JE. Anomalies of the coronary arteries. Report of four cases. *Arch Pathol.* 1948;45:766-71.
3. Alexander RW, Griffith GC. Anomalies of the coronary arteries and their significance. *Circulation.* 1956;14:800-5.
4. Taylor AJ, Rogan KM, Virmani R. Sudden cardiac death associated with isolated congenital coronary artery anomalies. *J Am Coll Cardiol.* 1992;20:640-7.
5. Roberts WC. Major anomalies of coronary arterial origin seen in adulthood. *Am Heart J.* 1986;111:941-63.
6. Frommelt PC, Frommelt MA. Congenital coronary artery anomalies. *Pediatr Clin North Am.* 2004;51:1273-88.
7. Reul RL, Cooley DA, Hallman GL, Reul GJ. Surgical treatment of coronary artery anomalies. *Tex Heart Inst J.* 2002;29:299-307.
8. Furukawa K, Itoh T. Direct coronary Reimplantation for repair of anomalous aortic origin of left or right coronary artery. *Ann Thorac Surg.* 2005;79:389-90.
9. Fernandes ED, Kadivar H, Hallman GL, Reul GJ, Ott DA, Cooley DA. Congenital malformations of the coronary arteries: the Texas Heart Institute experience. *Ann Thorac Surg.* 1992;54:732-40.
10. Mustafa I, Gula G, Radley-Smith R, Durrer S, Yacoub M. Anomalous origin of the left coronary artery from the anterior aortic sinus: a potential cause of sudden death. Anatomic characterization and surgical treatment. *J Thorac Cardiovasc Surg.* 1981;82:297-300.
11. Romp RL, Helong R, Landolfo CK, Sanders SP, Miller CE, Ungerleider RM, et al. Outcome of unroofing procedure for repair of anomalous aortic origin of left or right coronary artery. *Ann Thorac Surg.* 2003;76:589-96.
12. van Son JAM, Mohr FW. Modified unroofing procedure in anomalous aortic origin of left or right coronary artery. *Ann Thorac Surg.* 1997;64:568-9.
13. Roberts WC, Siegel RJ, Zipes DP. Origin of the right coronary artery from the left sinus of Valsalva and its functional consequences: analyses of 10 necropsy patients. *Am J Cardiol.* 1982;49:863-8.
14. Angelini P. Coronary artery anomalies—current clinical issues. *Tex Heart Inst J.* 2002;29:271-8.
15. Kaku B, Shimizu M, Yoshio H, Ino H, Mizuno S, Kanaya H, et al. Clinical features of prognosis of Japanese patients with anomalous origin of the coronary artery. *Jpn Circ J.* 1996;60:731-41.
16. Mishra S, Wolfram RM, Torguson R, Xue Z, Gevorkian N, Chu WW et al. Procedural results and outcomes after extensive stent coverage with drug-eluting stent implantation in single coronary lesions. *Am J Cardiol.* 2006;98:357-61.
17. Bauters C, Hubert E, Prat A, Bougrimi K, Van Belle, McFadden EP, et al. Predictors of restenosis after coronary stent implantation. *J Am Coll Cardiol.* 1998;31:1291-8.
18. Cohen AJ, Grishkin BA, Helsel RA, Head HD. Surgical therapy in the management of coronary anomalies: emphasis on utility of internal mammary artery grafts. *Ann Thorac Surg.* 1989;47:630-7.
19. Sabik JF, Lytle BW, Blackstone EH, Khan M, Houghtaling PL, Cosgrove DM. Does competitive flow reduce internal thoracic artery graft patency? *Ann Thorac Surg.* 2003;76:1490-7.
20. Berger A, McCarthy PA, Siebert U, Carlier S, Wijns W, Heyndrickx G, et al. Long-term patency of internal mammary artery bypass grafts relationship with preoperative severity of the native coronary artery stenosis. *Circulation.* 2004;110(suppl II):II36-40.
21. Navia D, Cosgrove DM, Lytle BW, Taylor PC, McCarthy PM, Stewart RW, et al. Is the internal thoracic artery the conduit of choice to replace a stenotic vein graft? *Ann Thorac Surg.* 1994;57:40-4.
22. Rogers OR, Leacche M, Mihaljevic T, Rawn JD, Byrne JG. Surgery for anomalous origin of the right coronary artery from the left aortic sinus. *Ann Thorac Surg.* 2004;78:1829-31.