

# Aortic Valve Replacement in 8 Adults with Anomalous Aortic Origin of Coronary Artery

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Key words: Aortic valve/ surgery; combined modality therapy; coronary vessel anomalies/complications/ diagnostic imaging; heart valve prosthesis implantation/methods; postoperative complications; retrospective studies; transcatheter aortic valve replacement; treatment outcome

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© 2019 by the Texas Heart® Institute, Houston Patients with anomalous origin of a coronary artery during aortic valve replacement (AVR) are at risk of coronary compromise. Large case series are lacking. In this retrospective study, we review our experience with this condition.

From August 2014 through June 2016, 8 adult patients (mean age,  $74 \pm 17.5$  yr; age range, 33-86 yr; 5 men) with anomalous aortic origin of a coronary artery underwent surgical or transcatheter AVR at our institution. Six patients had aortic stenosis; 2 had aortic insufficiency, one of whom had an associated aortic root aneurysm.

In 7 patients, the left anomalous coronary artery originated from the right aortic sinus, and in one, the right coronary artery arose from the left cusp. The anatomic course was revealed by means of 3-dimensional computed tomographic angiography. No patient underwent primary aortic reimplantation of the anomalous artery. Two had the artery mobilized from encircling the annulus too closely and then underwent coronary artery bypass grafting. Two high-risk patients underwent transcatheter AVR. After surgical AVR, 2 patients had perioperative myocardial infarction caused by coronary compression, so percutaneous coronary intervention was performed. One patient died of sepsis 3 months after discharge from the hospital.

In our patients, AVR sometimes led to severe perioperative complications. Successful AVR depended on 3-dimensional computed tomographic angiographic findings, individual anatomic variations, and appropriate treatment choices. **(Tex Heart Inst J 2019;46(3):189-94)** 

n estimated 0.2% of individuals who undergo angiography have some form of anomalous aortic origin of the coronary arteries (AOCA). Patients with AOCA who have aortic valve replacement (AVR) are at risk for perioperative myocardial ischemia, because the prosthetic valve can compromise the coronary arteries.<sup>1,2</sup>

Although AOCA was first identified in 1948,<sup>3</sup> only recently, through single case reports, has caution emerged about the dangers of AVR in patients with AOCA. They need individualized surgical treatment to minimize complications.<sup>4,5</sup> Because the combination of aortic stenosis and AOCA is rare, no consensus guidelines to standardize optimal surgical therapy have been established.<sup>6,7</sup>

We report our experience with 8 adult patients who had AOCA and underwent AVR, and we present insights into the pre- and intraoperative technical considerations that affect clinical outcomes, specifically the potentially fatal complication of perioperative myocardial ischemia.

# **Patients and Methods**

From August 2014 through June 2016 at our institution, 323 patients underwent surgical AVR; and 134, transcatheter AVR (TAVR). Among those patients, 8 had AOCA. Our institutional review board approved this retrospective review. Data collected from the patients' health records included pertinent clinical characteristics and comorbidities as defined by the Society of Thoracic Surgeons. Three-dimensional 64-slice computed tomographic reconstruction angiography (3D-MDCTA) of the heart, coronary angiography, and echocardiography had been used to evaluate aortic valves and coronary anatomy preoperatively.

After AOCA was identified angiographically, patients were evaluated for the danger of coronary compromise. Multimodal imaging was used to guide the intra- and postoperative treatment of patients with aortic stenosis, including: transthoracic echocardiography in color-flow Doppler mode, 2-dimensional computed tomography, and 3-dimensional reconstruction views of 64-slice computed tomograms. To definitively evaluate the coronary anatomy and its relationship to the aortic sinuses, the preferred imaging method was 3D-MDCTA anatomic reconstruction with contrast.<sup>8</sup>

## Results

Table I summarizes the clinical and perioperative characteristics of the 8 patients (5 men and 3 women; mean age,  $74 \pm 17.5$  yr; age range, 33-86 yr), as well as surgical details.

In 7 patients, as is typical in AOCA, the left circumflex coronary artery originated from the right aortic sinus and coursed posterior to the aortic annulus. In Patient 5, the anomalous right coronary artery originated from the left coronary cusp and coursed anterior to the aortic annulus.

Six patients underwent surgical AVR, and 2, TAVR. The mean hospital stay was 12.5 days (range, 2–30 d). Among the 6 surgical patients, 5 underwent open AVR with use of noneverting pledgeted sutures. The remaining surgical patient had aortic root replacement in which everting sutures were used to secure the prosthesis onto the aortic annulus. All prostheses were undersized except when the anomalous vessel was bypassed.

Patients 1 and 2 had postoperative myocardial infarction caused by compression of the anomalous vessel, and they underwent percutaneous coronary intervention (PCI). Postoperative coronary angiograms enabled expeditious identification and correction of the compression. Patients 3 and 4 underwent coronary artery bypass grafting (CABG) at the same operation.

Patients 7 and 8 were selected directly for TAVR on the basis of multiple comorbidities, including a patent left internal thoracic coronary artery graft in one.

There were no in-hospital deaths; a late death 3 months after hospital discharge (Patient 1) was related to sepsis and multiorgan failure.

### **Case Descriptions**

Operative outcomes depended on thorough evaluation and approach. After confirming AOCA, the surgeon needed to consider multiple options based on perioperative findings.

Patient 1 was our first experience with AVR complicated by AOCA. Her preoperative angiogram showed the anomalous coronary artery (Fig. 1A). She had myocardial ischemia, which appeared to be caused by coronary obstruction from a valve suture, near the junction between the left and noncoronary cusps (Fig. 1B). Periadventitial hemorrhage or operative trauma may also have caused compression. Initial postoperative clinical evaluation had not indicated the gravity of the problem; moreover, diagnosis was perhaps clouded by postoperative chest pain that the surgeon perceived to be incisional. In addition, this patient's paced rhythm might have made interpretation of the ST segments challenging. Her condition declined precipitously. Of the treatment options for acute myocardial ischemia caused by coronary artery compression, PCI without delay was best (Fig. 1C), because longer time between identifying myocardial ischemia and intervention may increase postoperative mortality rates.

Patient 1 underwent mitral valve replacement 6 weeks after discharge from the hospital (via a standard left atrial approach) to correct postoperative ischemic mitral regurgitation. A transseptal approach for her mitral valve replacement carried too much risk of crushing the previously placed stent during atrial exposure and surgical traction across the left atrial dome.

Patient 2 had paroxysmal atrial fibrillation; excising her left atrial appendage was not an option because its base was too close to the aberrant vessel. (In Patients 1 and 2, no benefit was associated with using direct exposure of the small AOCA vessel size for preventive grafting.) Preoperatively, current-generation MDCTA with 3-dimensional volume-rendering was sufficient for evaluation in Patient 2 (Fig. 2A). However, reliance on postoperative CTA risked the inability to detect a compressed, malperfused, or nonenhanced aberrant vessel. Coronary angiography provided greater accuracy (Fig. 2B–C).

In Patient 3 (age, 33 yr), no AOCA had been suspected; however, preoperative 3D-MDCTA revealed the anomalous vessel (Fig. 3). With use of cardiopulmonary bypass, vessel mobilization was attempted and then abandoned, because the vessel was adhered to the surrounding tissue. The most notable feature of this patient's coronary anatomy was that, after the aortotomy, the anomalous vessel orifice was found to be slit-like and too close to the fused raphe. Therefore, the patient underwent CABG.

In Patient 4, the anomalous vessel was difficult to reach because of its deep intramyocardial course and its location between the pulmonary artery, the left atrial appendage, and the nearby left anterior descending coronary artery (Fig. 4A). It would not have been possible to free this vessel easily or completely in preparation for bypass without transecting the aorta, which was done (Fig. 4B). Venous bleeding after cardiopulmonary bypass had been discontinued probably originated from multiple small distal branches of the coronary sinus. This bleeding was difficult to control, so the aorta was cross-clamped and the heart arrested again.

Variable	Patient Number							
	1	2	3	4	5*	6	7	8
Age (yr), sex	80, F	86, F	33, M	81, M	67, M	81, M	79, F	85, M
Body mass index (kg/m²)	21.45	26.67	28	26.68	37.69	32	22.51	24.76
Body surface area (m²)	1.52	1.8	2.14	1.91	2.56	2.16	1.56	1.88
Diabetes mellitus	No	No	No	No	Yes	No	Yes	No
Hypertension	Yes	Yes	No	Yes	Yes	No	Yes	No
Hyperlipidemia	Yes	Yes	No	Yes	Yes	No	Yes	No
Hypothyroidism	No	Yes	No	No	No	No	No	No
Smoking history	Yes	Yes	Yes	Yes	No	No	Yes	Yes
COPD	No	No	No	No	No	No	Yes	Yes
Cancer	No	No	No	No	No	No	Yes	No
Aortic stenosis	No	No	No	Yes	No	Yes	Yes	Yes
Bicuspid aortic valve	Yes	Yes	No	No	No	No	No	No
Previous stroke or PVD	No	Yes	No	Yes	No	No	Yes	Yes
Previous operations	None	None	None	None	None	None	Patent LITA graft	MVR
Operative evaluation								
Aortic insufficiency	No	No	Yes	No	Yes	No	No	No
Atrial fibrillation	No	Yes	No	No	No	No	No	Yes
Heart failure	No	No	Yes	No	Yes	No	No	Yes
LVEF	0.65	0.70	0.25	0.70	0.60	0.76	0.55	0.55
Mean PAP >25 mmHg	No	Yes	Yes	No	No	No	No	No
STS risk score (%)	NA	NA	NA	NA	NA	NA	9	10
Operative details								
AVR method	Surgical	Surgical	Surgical	Surgical	Surgical	Surgical	Trans	Trans
Prosthesis type, size (mm)	Perimount Magna, 19ª	Trifecta, 21 <sup>b</sup>	Perimount Magna, 27	Perimount, 21	Valsalva, 25 <sup>c</sup>	Trifecta, 21	Sapien 3, 23 <sup>a</sup>	Sapien 3 29
Concomitant procedure	None	None	CABG	CABG	Ao RR	None	None	None
CPB/CC time (min)	113/82	141/92	152/128	300/228	260/192	212/139	None	None
Postoperative complications	MI	MI	None	None	None	None	None	None
Length of stay (d)	30	11	5	11	14	15	2	2

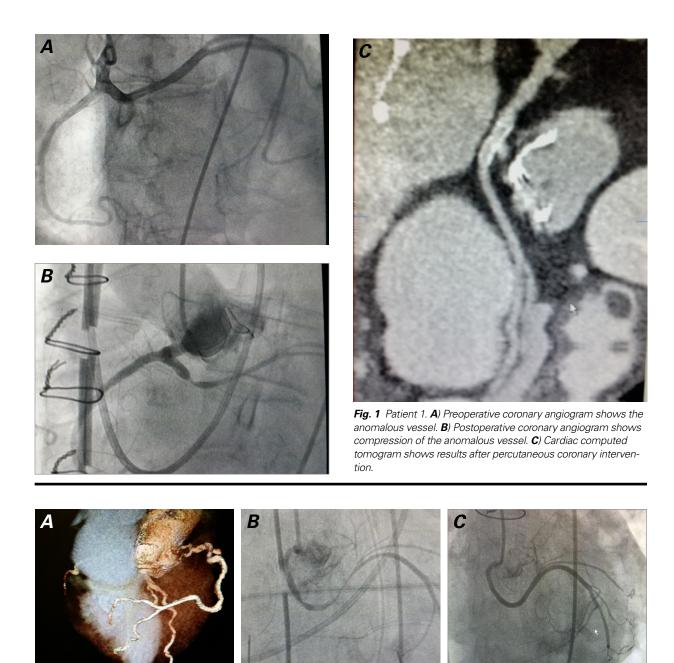
Ao RR = aortic root replacement; AVR = aortic valve replacement; CABG = coronary artery bypass grafting; CC = aortic cross-clamp; COPD = chronic obstructive pulmonary disease; CPB = cardiopulmonary bypass; F = female; LITA = left internal thoracic artery; LVEF = left ventricular ejection fraction; M = male; MI = myocardial infarction; MVR = mitral valve replacement; NA = not available; PAP = pulmonary artery pressure; PVD = peripheral vascular disease; STS = Society of Thoracic Surgeons; Trans = transcatheter

\*Patient 5 had an anomalous right coronary artery; all others had an anomalous left circumflex coronary artery.

<sup>a</sup> Edwards Lifesciences Corporation

<sup>b</sup> St. Jude Medical, part of Abbott

<sup>c</sup> Vascutek, part of Terumo Cardiovascular Systems



**Fig. 2** Patient 2. **A**) Preoperative computed tomographic angiogram (3-dimensional reconstruction) shows an anomalous vessel. Coronary angiograms show **B**) immediate postoperative narrowing from compression of the coronary artery by the prosthetic aortic valve, and **C**) the widely patent artery after percutaneous coronary intervention.

In Patient 5, separating the origins of the coronary arteries from the aorta was not difficult, because the right coronary artery originated from the left coronary cusp and coursed anterior to the aortic root. In this case, the choice of reimplanting only the left ostium was influenced by the surgeon's decision to avoid kinking. Despite some calcium around the origin of the anomalous coronary artery, the left coronary button was implanted easily onto the composite graft. The patient's 3 valve leaflets were flimsy and had several fenestrations, so a valve-sparing procedure could not be performed.

In Patient 6, mobilizing the anomalous vessel was not possible because of its small size. However, to avoid impinging on the anomalous coronary artery, the sutures were placed low below the aortic annulus, and the prosthesis was undersized. Despite these precautions, the procedure resulted in compression of the anomalous vessel (Fig. 5).



*Fig. 3* Patient 3. Preoperative computed tomographic angiogram (3-dimensional reconstruction) shows an unexpected anomalous vessel.

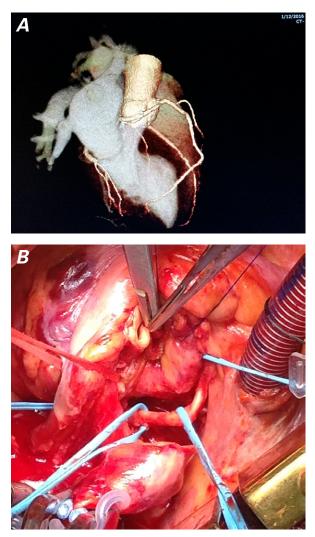
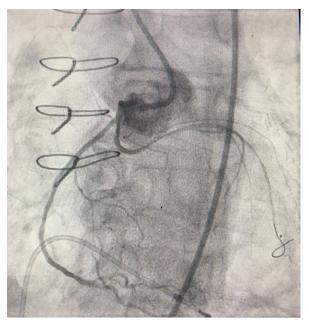


Fig. 4 Patient 4. A) Preoperative computed tomographic angiogram (3-dimensional reconstruction) shows the anomalous vessel. B) Intraoperative photograph shows the aorta completely transected for access to the vessel.



**Fig. 5** Patient 6. Postoperative coronary angiogram shows narrowing caused by compression of the coronary artery by the prosthetic aortic valve.

Patients 7 and 8 were at high surgical risk, so TAVR, although more technically challenging than open AVR, was the minimally invasive solution of choice. Using the SAPIEN S3<sup>®</sup> Transcatheter Heart Valve (Edwards Lifesciences Corporation) eased engaging the coronary ostia, because this stent is not as tall as other TAVR stents. Although surgical sutureless AVR and TAVR prostheses have been proposed in the management of inoperable surgical patients with severe aortic stenosis, the long-term results will need to be determined.<sup>9,10</sup>

## Discussion

No evidence-based practice guidelines have been developed for treating patients who have AOCA and need AVR. On the basis of our experience in 8 patients, multiple distinct perioperative interventions were helpful; they may have minimized associated risks and improved clinical outcomes. We propose the following strategies for treating AVR in patients with AOCA.

First, the consensus statement in the American College of Cardiology/American Heart Association 2014 guidelines for the management of patients with valvular heart disease does not specify the role of 3D-MDCTA in patients undergoing AVR.<sup>11</sup> We recommend this imaging method for preoperative evaluation of the course and size of anomalous coronary arteries.

Second, performing AVR in the hybrid operating room (if available) is recommended so that coronary angiograms can be obtained without delay, as has been suggested.<sup>12</sup>

Third, some investigators have convincingly argued that in patients with AOCA who need AVR, CABG is acceptable when anomalous vessels are large enough.<sup>13</sup> This may also include vessel mobilization away from the aortic root. Complementary endovascular therapy, including PCI with or without stenting, is also an option to relieve cardiac ischemia; TAVR may be considered when prohibitive surgical risks exist.

Fourth, we recommend using coronary angiography to identify potential AOCA before performing any AVR, especially in patients younger than 45 years of age who may not have angina.

We conclude from our small series that no "one-sizefits-all" approach will reduce AVR-related vascular compromise in patients with AOCA.

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