Setting things “right”: right internal mammary artery on anomalous right coronary artery - a case report

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Abstract
Anomalous aortic origin of a coronary artery (AAOCA) is a rare pathology that may cause episodic ischemia owing to possible vessel compression during systolic expansion of the aortic root. This anomaly can lead to myocardial infarction, malignant arrhythmias and sudden cardiac death (SCD). Several surgical techniques have been described; however, there are no defined guidelines regarding the treatment of AAOCA. We report the case of a 47-year-old woman with ectopic origin of the right coronary artery (RCA) from the left sinus of Valsalva, with an inter-arterial course of the proximal segment of the artery, running between the aorta and the pulmonary trunk. Revascularization was accomplished by harvesting the right internal mammary artery (RIMA) and anastomosing it to the anomalous RCA, given the small portion of the RCA following an intramural course and our familiarity with the procedure. The RCA was ligated proximal to the anastomosis to avoid the string sign phenomenon. This procedure is safe and fast and can be considered an alternative to coronary reconstruction.

Keywords
Anomalous coronary artery, congenital heart disease, cardiac surgery, coronary artery bypass graft, internal mammary artery, aorta

Date received: 29 June 2021; accepted: 30 September 2021

Introduction
Anomalous aortic origin of a coronary artery (AAOCA) occurs in 1% of the general population and is reported to cause 5% to 35% of sudden deaths in the young and almost 19% of sudden deaths in young...
The prevalence of a right coronary artery (RCA) originating from the left sinus of Valsalva varies in the literature but is estimated to be 0.02% to 0.9%. An ectopic RCA can have different possible courses, which may be clinically benign or malignant: 1) retrocardiac, or RCA running in the posterior atrioventricular groove; 2) retroaortic; 3) interarterial, between the aorta and the pulmonary artery; 4) intraseptal; and 5) precardiac, anterior to the right ventricular outflow tract (RVOT).

Rigatelli et al. classified AAOCA into five categories according to the clinical significance, which depends on the route of the coronary artery relative to the great vessels and the presence of concomitant coronary artery disease, offering a new tool in decision-making for both surgical and interventional management. According to the aforementioned classification (Table 1), an ectopic origin of the RCA from the left sinus of Valsalva has Class III clinical relevance (severe). This is because an ectopic origin of the RCA from the left sinus of Valsalva with the artery running between the aorta and pulmonary trunk may cause episodic ischemia owing to possible vessel compression during systolic expansion of the aortic root, leading to myocardial infarction and sudden cardiac death (SCD). Moreover, this anomaly is associated with a higher risk of malignant ventricular arrhythmias.

We present the case and our treatment approach for a 47-year-old woman with an anomalous RCA originating from the left sinus of Valsalva that followed an interarterial course.

**Case report**

A 47-year-old woman was referred to our hospital for recurrent palpitations during exercise, with worsening symptoms in the previous 3 months. Her medical history was unremarkable except for transient, episodic chest pain. She was asymptomatic at rest.

| Table 1. Rigatelli classification. |
|-------------------------------|
| **I-Benign**                  | • Ectopic origin of the LCx from the RS |
|                               | • Separate origin of the LCx and LAD |
|                               | • Ectopic origin of the LCx from the RCA |
|                               | • Ectopic coronary origin from the AO |
|                               | • Dual LAD type I to IV |
|                               | • Myocardial bridge (score: ≤5) |
|                               | • Intercoronary circulation |
| **II-Relevant**               | • Coronary artery fistula |
|                               | • Single coronary artery R/L, I/II/III, A/P |
|                               | • Ectopic origin of the LCA from the PA |
|                               | • Atretic coronary artery |
|                               | • Hypoplastic coronary artery |
| **III-Severe**                | • Ectopic origin of the LCA from the RS |
|                               | • Ectopic origin of the RCA form the LS |
|                               | • Ectopic origin of the RCA from the PA |
|                               | • Single coronary artery R/L, I/II/III B |
|                               | • Myocardial bridge (score: 5) |
| **IV-Critical**               | • Class II and superimposed CAD |
|                               | • Class III and superimposed CAD |

AO, ascending aorta; CAD, coronary artery disease; L, left; LAD, left anterior descending coronary artery; LCA, left coronary artery; LCx, left circumflex coronary artery; LS, left sinus; PA, pulmonary artery; R, right; RCA, right coronary artery; RS, right sinus.
pregnancy-related hyperthyroidism and a smoking habit. Her physical examination findings were also unremarkable, with normal blood pressure, normal jugular venous pressure, and normal heart sounds, while her resting electrocardiogram (ECG) showed sinus rhythm with isolated premature ventricular contractions (PVCs). Echocardiography demonstrated absence of valvular disease, preserved ejection fraction (EF: 66%), and mild hypokinesis of the basal and medial segments of the left ventricle.

Cardiac computed tomography (CT) was performed and revealed an ectopic origin of the RCA from the left sinus of Valsalva, with an interarterial course of the proximal segment of the artery, running between the aorta and the pulmonary trunk [Figure 1 and Figure 2]. The posterior descending artery and the posterolateral artery originated from the RCA. There were no atherosclerotic lesions in either the RCA or the left coronary artery (LCA). CT also revealed the presence of a 12-mm lung nodule.

We performed coronary artery bypass grafting (CABG) and concomitant excision of the lung nodule, which proved to be a hamartoma on histological examination. The procedure was performed under cardiopulmonary bypass with aortic cross-clamping and the administration of antegrade cold blood cardioplegia. The right internal mammary artery (RIMA) was harvested, skeletonized, and anastomosed in situ to the RCA. Subsequently, the RCA was ligated proximal to the anastomosis to prevent coronary steal syndrome and flow competition [Figure 3].

The patient had an uneventful postoperative course and was discharged home on the 8th postoperative day.

Resting 12-lead ECG, 24-hour Holter monitoring, and exercise ECG (stress test) 3, 6, and 12 months after discharge showed sinus rhythm, and no PVCs were observed.

Figure 1. Preoperative heart CT scan. The CT image shows the origin of the RCA from the left coronary sinus with an interarterial course (arrow). CT, computed tomography; AO, aorta; LCA, left coronary artery; RCA, right coronary artery.

Figure 2. Preoperative 3D CT reconstruction showing the origin and interarterial course of the RCA in our 47-year-old patient. 3D, three-dimensional; CT, computed tomography; AO, aorta; PA, pulmonary artery; LCA, left coronary artery; LAD, left anterior descending artery; CX, circumflex artery; RCA, right coronary artery.
Follow-up echocardiography revealed preserved EF and no wall motion abnormalities. CT re-evaluation 3 months after surgery revealed patency of the RIMA graft.

The reporting of this study conforms to the CARE guidelines.8

Discussion

SCD is frequently reported in the literature as the first and only presentation of AAOCA in patients with an interarterial course. Considering this, as well as the hypokinesis of the left ventricle, which was suggestive of ischemia, we decided to treat our patient even though she did not present with life-limiting symptomatology.

Embryologically, the coronary circulation originates from proliferation of endothelial cells, vascular smooth muscle cells, and fibroblasts of the capillary plexus, which migrate into the aortic sinuses. These cells penetrate the aortic wall from a capillary ring surrounding the aortic root, giving origin to the coronary circulation. In AAOCA, the cells of the capillary plexus surrounding the aorta fail to reach and/or penetrate the two normal sites on the aorta, the right and left coronary sinus, probably due to vascular endothelial growth factor C (VEGF-C) deficiency.9

The pathophysiology of SCD is not completely understood; however, a few mechanisms have been proposed.10–14 In patients with an interarterial course, there can be vascular compression of the proximal segment of the RCA, which passes between the aorta and pulmonary artery, because of engorgement of the two vessels during systole. This may reduce blood supply to the myocardium, especially to the right ventricle, causing ischemia and leading to arrhythmias that may have a fatal outcome. Another proposed mechanism may be attributed to the intramural course of the RCA, with possible lumen diameter reduction during systole, secondary to myocardial contraction. An additional mechanism is represented by the angulation of the artery, with proximal stenosis occasionally observed at the origin of the RCA, especially in the presence of a retrocardiac course. In pediatric patients, diagnosis is generally performed through echocardiography, even if CT is necessary to further investigate the coronary tree, especially in patients weighing more than 40 kg. The use of Doppler ultrasonography improves the precision of echocardiographic diagnosis because flow velocity is lower in diastole, whereas systolic compression reduces flow. In adolescents or adults, structural magnetic resonance imaging (s-MRI) is the ideal imaging tool because it allows precise and reliable diagnosis of

Figure 3. Postoperative 3D CT reconstruction
The right internal mammary artery is anastomosed to the mid-RCA, which is ligated proximally. Postoperative heart CT showing the RIMA grafted on the RCA.

3D, three-dimensional; CT, computed tomography; AO, aorta; PA, pulmonary artery; RIMA, right internal mammary artery; RCA, right coronary artery; LAD, left anterior descending artery.
high-risk coronary artery anomalies, and this imaging modality is readily available in most centers. Cardiac CT (CCT) also offers high-quality imaging. However, intramural aortic course evaluation may be challenging, and exact quantification of stenosis cannot be achieved by s-MRI or CCT. Only cross-sectional imaging perpendicular to the vessel’s direction obtained during both cardiac phases can help quantify the area of stenosis. This imaging can be achieved only with catheter angiography, which is used for definitive imaging of the vessel and stenosis quantification, combined with intravascular ultrasonography (IVUS) evaluation.

Several surgical techniques have been proposed to repair AAOCA. However, no comparison between techniques has been reported; therefore, there is no evidence supporting superiority regarding mortality, safety, and long-term outcomes among the described procedures. Unroofing the RCA to treat this anomaly is often proposed in the literature, which provides excellent midterm results, and is mostly used when the intramural length of the anomalous vessel is conspicuous. Another applicable technique is reimplantation of the RCA, which consists of dissecting and mobilizing the proximal portion of the RCA, transecting the artery after aortic cross-clamping, and reimplanting the RCA into the non-coronary sinus. Other techniques, not applicable to our patient, are ostioplasty, which is the creation of a neo-ostium from the normal ostium to the ectopic artery, with pericardial patch augmentation of the vascular wall, as well as percutaneous coronary intervention (PCI) stenting. Another technique is pulmonary artery translocation, where the distal main pulmonary artery is transected and then relocated to the left pulmonary artery to prevent compression of the anomalous coronary artery. Alternatively, transection of the right pulmonary artery at its origin and anastomosis to the pulmonary trunk anterior to the aorta, with patch augmentation, can be performed. Each strategy has pitfalls, and the choice should be based on individual patient anatomy and the center’s surgical experience. For example, possible pitfalls of coronary unroofing are the possibility of coronary injury and, in almost 15% of the cases, the necessity of aortic valve resuspension, with the consequent risk of aortic regurgitation, albeit minimal. In comparison, one of the most important concerns regarding the revascularization strategy is ligating the coronary artery because the flow in the anastomosed mammary artery in the early stages may not adequately sustain myocardial perfusion of the right ventricle (RV). This is because there are no actual anatomical stenoses in the native coronary artery; therefore, some authors limit CABG to patients with concomitant coronary artery disease or as a bailout procedure for failed anatomical repair. Because AAOCA is rare, not all centers have adequate experience with all of the aforementioned techniques. However, surgical risk in published series is very low, with excellent intermediate-term survival despite concerns about the long-term impact of coronary surgery in pediatric patients related to scarring or accelerated atherosclerosis.

In our patient, revascularization was accomplished by harvesting the RIMA and anastomosing it to the anomalous RCA, as reported by Reul et al. This approach was chosen because the patient was an adult, and RIMA dimensions were adequate; because of the small portion of the RCA following an intramural course; and finally, because of our familiarity with the procedure. The RCA was subsequently ligated proximal to the anastomosis, as described by Gaudino et al. to avoid the string sign phenomenon. Competitive flow predisposes the internal mammary arteries to the string sign phenomenon,
which is especially prevalent in revasculariza-
tion of the right coronary artery when the
vessel is not severely stenosed or completely
occluded. This is why we strongly suggest
ligating the coronary artery when using the
aforementioned method, even though this is an
admittedly bold choice, as it removes the “safety
net” represented by native coronary flow with a
failed anastomosis.

There are no defined guidelines regarding
the treatment of AAOCA. The clinical
indication for intervention is based on the
calculated risk of SCD, considering both
the patient’s anatomical features and clinical
presentation, as well as the patient’s
associated comorbidities. However, an ini-
tial algorithm for the treatment of anom-
ylies of the coronary arteries has been
proposed in association with the Rigatelli
classification, where Class I, or benign cor-
donary artery anomalies, may not require
treatment, Class II anomalies require care-
ful follow-up, and Class III and IV generally
require surgical correction, as stated in
the consensus statement in the Guidelines
for Management of Adults with
Congenital Heart Disease.30 Age is an
important determinant for treatment
choice because pediatric patients aged 10
years or older are referred for surgical inter-
vention owing to the increased risk of SCD
during exercise.31,32

Follow-up for AAOCA patients depends
on the chosen management plan, even
though all patients must receive lifelong
cardiac check-ups. Close periodic evalua-
tion in patients who do not undergo surgi-
cal correction of the anomaly is of utmost
importance, while in surgical patients, the
time between each cardiac assessment
may be extended after the first year.
Generally, complete follow-up evaluation
requires echocardiography, ECG at rest, a
stress test, and 24-hour Holter ECG
monitoring.

Conclusion
AAOCA represents a rare pathology,
which, especially in the adult population,
can prove to be a therapeutic challenge.
Consensus on AAOCA treatment is limited,
likely owing to its rarity; however, it is
widely accepted that patients with clinically
malignant anomalies must undergo surgical
treatment to avoid catastrophic complica-
tions secondary to ischemic events. While
techniques such as coronary unroofing,
ostium reimplantation, or neo-ostium cre-
ation are valid options, in adults, CABG is a
fast, effective, and safe strategy. Using
CABG requires creating an occlusion prox-
imal to the anastomosis, to avoid the string
sign phenomenon. Careful consideration of
the treatment approach is fundamental to
choose the correct technique for an individ-
ual patient, as each technique requires a tai-
lored methodology.

Ethics statement
This study was a descriptive case report, and
ethics committee approval was not required.
The patient participating in this study provided
written informed consent.

Declaration of conflicting interest
The authors declare that there is no conflict of
interest.

Funding
This research received no specific grant from any
funding agency in the public, commercial, or
not-for-profit sectors.

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