Evaluation of Anomalous Aortic Origins of the Coronaries by 64-Slice Cardiac Computed Tomography

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Approximately 20% of coronary artery anomalies produce sudden death or life-threatening symptoms, including arrhythmias, syncope, and myocardial infarction. The most common clinical symptom of coronary artery anomaly is angina or exertional syncope. Physical examination is usually unrevealing in the absence of myocardial infarction or symptoms of ongoing ischemia. The rapid advent of cardiac computed tomography (CT) technology has made it an important adjunct to the diagnosis of coronary anomalies by angiography. The authors describe the case of a 54-year-old white man who presented with gangrenous toes. He had severe peripheral vascular disease, a femoral-popliteal bypass graft, residual hemiparesis from an ischemic stroke, hypertension, deep vein thrombosis, and a recent myocardial infarction. He underwent a 64-slice cardiac CT angiogram, which showed an interarterial course of the left main coronary artery between the aorta and the pulmonary trunk.
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Key words: Computed tomography • Anomalous aortic origins of the coronaries • Left circumflex artery • Right anterior oblique • Magnetic resonance angiography

Congenital anomalies of the coronary arteries occur in 0.2% to 1.2% of the general population.[1-3] Most coronary artery anomalies are not clinically symptomatic and are found incidentally during angiographic evaluation for other cardiac diseases. Approximately 20% of coronary artery anomalies produce sudden death or life-threatening symptoms, including arrhythmias, syncope, and myocardial infarction. Congenital coronary artery anomalies are the second most common cause of sudden death due to structural heart disease in young athletes.[4]

The rapid advent of cardiac computed tomography (CT) technology has made it an important adjunct to the diagnosis of coronary anomalies by angiography. We discuss a patient with an anomalous left main (LM) coronary artery that was suspected during coronary angiography and confirmed by 64-slice CT scan.

Case Presentation

A 54-year-old white man with severe peripheral vascular disease, a femoral-popliteal bypass graft, residual hemiparesis from an ischemic stroke, hypertension, deep vein thrombosis, and a recent myocardial infarction presented with gangrenous toes. A physical examination revealed nothing remarkable except for nonhealing arterial ulcers associated with the dry gangrene. An electrocardiogram showed T-wave inversions in the inferior leads, with an incomplete right bundle branch block pattern. A 2-dimensional echocardiogram showed a dilated left ventricle with inferoapical akinesis.

Further examination revealed that the patient had an occluded lower extremity bypass graft. He underwent stress myocardial perfusion imaging as part of the preoperative cardiac risk assessment. A dipyridamole sestamibi stress test showed an inferior and inferolateral wall infarct with no ischemia. The patient's left ventricular ejection fraction was 17%. He then underwent a left heart catheterization to evaluate his coronaries. It showed an anomalous circulation, with the LM coronary artery arising from the right coronary artery (RCA) (Figures 1 and 2). He subsequently underwent a 64-slice cardiac CT angiogram, which showed an interarterial course of the LM coronary artery between the aorta and the pulmonary trunk (Figures 3 and 4).

Although cardiothoracic surgery was considered, the patient was deemed to be at high risk for complications. Sudden death is usually associated with young age and vigorous exercise. The patient's age and nonambulatory status, coupled with his lack of symptoms, put him at low risk for sudden death. Hence, it was decided not to pursue surgical correction of his anomalous coronary artery. He underwent amputation of his gangrenous leg without any complications. He was advised to avoid heavy exercise.

Figure 1. *Coronary angiogram showing an empty left coronary sinus.* www.medreviews.com

Figure 2. *Coronary angiogram showing the left main artery originating from the right coronary artery.* www.medreviews.com

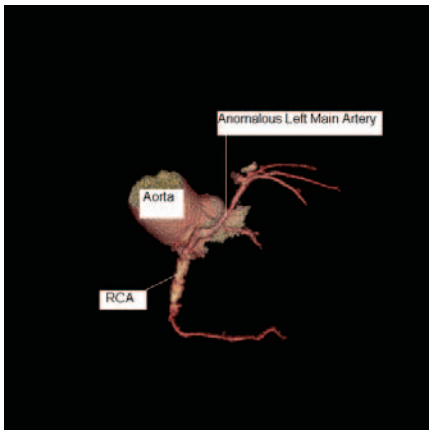

Figure 3. *This image from 64-slice cardiac computed tomography shows an anomalous left main coronary artery arising from the right coronary artery (RCA).* www.medreviews.com

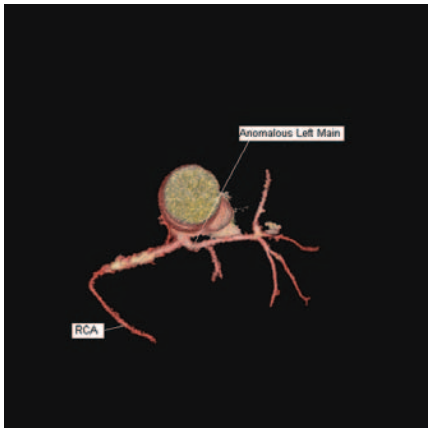

Figure 4. *Cardiac computed tomography (volume rendering) showing the anomalous left main artery and its branches. RCA, right coronary artery.* www.medreviews.com

Embryology

In early fetal development, the myocardium is nourished through sinusoids that develop to form the primitive coronary artery network. The definitive coronary artery system is established by endothelial buds that arise from the base of the truncus arteriosus and later grow and join these sinusoids.[5] Abnormal involution, bud position, or septation of the truncus arteriosus may lead to an abnormal origin of the coronary arteries.[6] These conditions may arise from the normal sinuses of Valsalva in the aorta or from the pulmonary artery.

Table 1
STS Congenital Heart Surgery Database Committee Nomenclature for Coronary Artery Anomalies

Anomalous pulmonary origins of the coronaries (APOC)
Anomalous aortic origins of the coronaries (AAOC)
Congenital atresia of the left main coronary artery (CALM)
Coronary arteriovenous fistulae (CAVF)
Coronary artery bridging (CB)
Coronary artery aneurysms (CAn)
Coronary stenosis

STS, Society of Thoracic Surgeons.

Clinical Presentation

The most common clinical symptom of coronary artery anomaly is angina or exertional syncope. Unfortunately, a patient may first present with sudden death. In a registry of sudden death in 286 competitive athletes younger than age 35 in whom cardiovascular disease was shown to be the cause of death at autopsy, an anomalous coronary artery of wrong sinus origin was responsible for 13% of cases, which was second in frequency to hypertrophic cardiomyopathy.[4] Physical examination is usually unrevealing in the absence of myocardial infarction or symptoms of ongoing ischemia.[7,8]

Types of Anomalies

A consensus report by the Society of Thoracic Surgeons Congenital Heart Surgery Database Committee outlined the various types of coronary artery anomalies (Table 1).[9] Of these, the 3 most common types of clinically significant anomalies that are treated surgically are coronary arteriovenous fistulae, anomalous pulmonary origins of the coronaries, and anomalous aortic origins of the coronaries (AAOC).[9] In the AAOC group, the 4 common subtypes are a left circumflex artery (LCx) arising from the right sinus of Valsalva or from the RCA, a single coronary artery from the left sinus of Valsalva, both coronary arteries from the right sinus of Valsalva, and an LM coronary artery or left anterior descending artery from the right sinus of Valsalva or from the RCA. Figures 5 through 9 illustrate some types of AAOC.

Complications

Complications depend on the site of origin of the anomalous artery and its further course as it supplies the myocardium. Origin of the LCx from the RCA is usually of no clinical significance due to its posterior course to the left ventricle. With the other types of origins, it is important to differentiate an interarterial course, in which the anomalous vessel courses directly between the aorta and the main pulmonary artery, from an intraseptal course, which may appear similar but in which the anomalous vessel passes more inferiorly within the muscular septum. The latter condition is associated with a comparatively benign course.[10]

An anomalous interarterial vessel may be associated with a poorer outcome.[8,10] A left coronary artery originating from the right sinus or an RCA arising from the left sinus can be interarterial. The interarterial course between the pulmonary artery and aorta may result in compression of the vessel, myocardial ischemia, or sudden death.[8,11,12] These complications usually occur during or immediately after exercise, and are more likely when an anomalous vessel supplies the left coronary artery distribution. Exercise causes expansion of the aortic root and pulmonary trunk, which leads to external compression of the coronary artery. It may also increase the preexisting angulation of the coronary artery takeoff, which results in a reduction of the luminal diameter in the proximal portion of the coronary artery and, therefore, predisposes the patient to ischemia.[8]

Diagnosis

X-Ray Coronary Angiography

X-ray coronary angiography has traditionally been considered the gold standard for diagnosing coronary artery anomalies. The most critical anomaly is the LM coronary artery arising from the right coronary cusp. The LM coronary artery may then take 1 of 4 courses: interarterial, retroaortic, septal, or through the anterior free wall of the right ventricle.

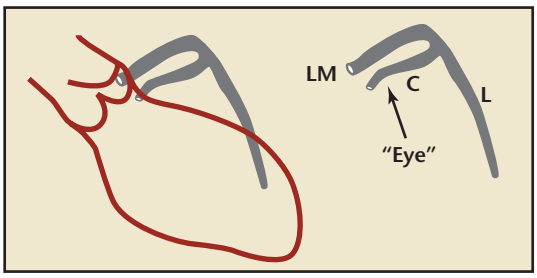

Figure 5. *Diagrammatic representation of an anterior course of an anomalous left coronary artery. LM, left main artery; C, circumflex artery; L, left anterior descending artery. Reproduced with permission from Kern MJ.*[14] www.medreviews.com

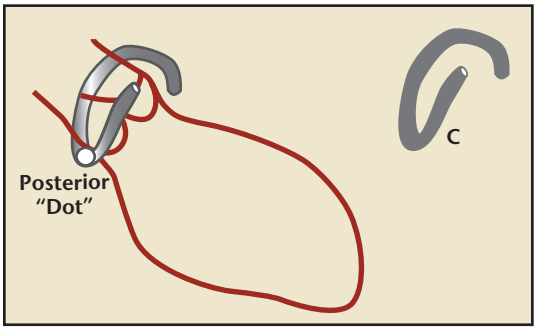

Figure 6. *Diagrammatic representation of a retroaortic course of a circumflex artery from the left coronary cusp. C, circumflex artery. Reproduced with permission from Kern MJ.*[14]
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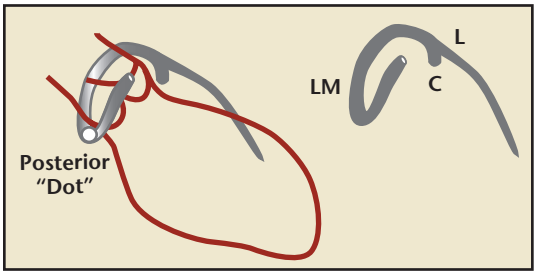

Figure 7. *Diagrammatic representation of a retroaortic course of the left main artery. LM, left main artery; C, circumflex artery; L, left anterior descending artery. Reproduced with permission from Kern MJ.*[14]
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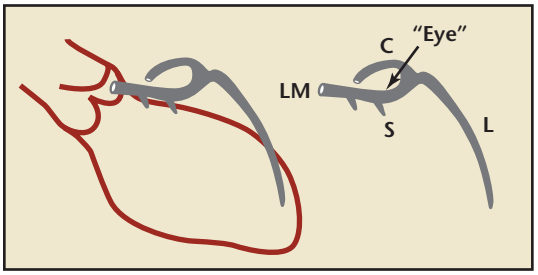

Figure 8. *Diagrammatic representation of a septal course of an anomalous left main artery. LM, left main artery; S, septals; C, circumflex artery; L, left anterior descending artery Reproduced with permission from Kern MJ.*[14] www.medreviews.com

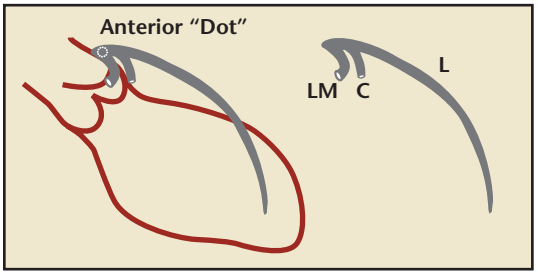

Figure 9. *Diagrammatic representation of an interarterial course of the left main artery. LM, left main artery; C, circumflex artery; L, left anterior descending artery. Reproduced with permission from Kern MJ.*[14]
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The interarterial course is associated with the worst prognosis, whereas the others are considered benign. This distinction makes accurate differentiation important.

In the view from the right anterior oblique, the LM coronary artery is seen "on-end" anterior to the aorta when the course is interarterial (Figure 9), and posterior (Figure 7) to the aorta when the course is retroaortic. When they take a septal course, the LM and LCx form an ellipse (or an "eye") to the left of the aorta, with the LM forming the inferior portion and the LCx forming the superior limb of the ellipse (Figure 4). In the anterior free wall of the right ventricle course, the LM and LCx form an ellipse to the left of the aorta, with the LM forming the superior portion and the LCx forming the inferior limb of the ellipse (Figure 5). An anomalous RCA arising from the left sinus of Valsalva that courses between the aorta and the pulmonary artery is seen "on-end" anterior to the aorta in the right anterior oblique view. This course has also been associated with myocardial ischemia.[13] The most common anomaly is the LCx arising from the proximal RCA. The LCx can also arise from the right coronary cusp.[14]

Such 2-dimensional projectional views, however, make delineation of the exact anatomic course of the anomalous vessels difficult. When x-ray angiography is compared with magnetic resonance angiography (MRA) or CT, the latter techniques seem better at delineating the origin, proximal course, and relationship with the surrounding structures.[15-19]

Echocardiography

Two-dimensional echocardiography is another option for the diagnosis of coronary artery anomalies. It is a noninvasive, safe, and attractive option because it provides good anatomic definition of the ostium and the proximal course of the coronary arteries. In a study examining coronary artery anatomy in 2388 asymptomatic children, adolescents, and young adults (up to age 21), 4 anomalies were identified by transthoracic echocardiography. There were 2 anomalous left coronary arteries and 2 anomalous RCAs.[20] A modified parasternal short axis view at the level of the aortic root was used to identify the coronary arteries. Color flow Doppler with the Nyquist limit lowered to a range of 20 to 40 cm/s seems to be ideal to visualize flow in the coronary arteries, at least in pediatric patients.[21] Less than 2% of the population studied had poor acoustic

windows that precluded accurate visualization of the coronary arteries.[20]

Echocardiographic screening for coronary artery anomalies is also feasible. In a prospective study of 1360 competitive athletes (ages 13 to 49), transthoracic echocardiography was able to accurately visualize the proximal epicardial coronary arteries in 93% of the study group. Twenty-eight athletes had variants of normal coronary arteries.[22] In another study of 3650 athletes, 96% had good windows, and coronary variants were found in 1.6%. Transthoracic echocardiography also accurately identified an interarterial course of a left coronary artery. This variant sometimes cannot be accurately identified even by coronary angiography.[23-25] However, it has been proposed that visualization of coronaries may be easier in athletes due to their favorable chest conformation, increase in coronary artery size as a result of training, and prolonged diastolic time due to bradycardia.[22,26,27]

In a study of 9 nonathletic patients with coronary artery anomalies diagnosed at cardiac catheterization, 7 had an interarterial course. The proximal courses of the coronary arteries were equivocal when viewed with angiography. However, the interarterial course and the relationship of the coronary artery with the great vessels were confirmed by transesophageal echocardiography.[28] Thus, both transthoracic and transesophageal echocardiography have been shown to identify coronary anomalies in both the pediatric and adult populations. However, the true positive and negative predictive value of echocardiography in identifying coronary artery anomalies remains unknown. Because it is relatively easy and noninvasive, routine evaluation of coronary arteries should become a standard part of all echocardiographic evaluations, especially during pre-athletic screenings.

Magnetic Resonance Angiography

MRA and cardiac CT have comparable results in identifying anomalous coronary arteries. In a blinded study of 38 patients, 19 of whom had known anomalies by coronary angiography, MRA was successful in identifying the origin and course of the anomalous vessel in all patients, even when the results of coronary angiography were equivocal.[17] Other studies have found similar results.[29,30] However, there are some limitations with MRA. The spatial resolution of MRA is lower than that of the latest CT scanners. Also, MRA imaging cannot be used in patients with pacemakers, defibrillators, or claustrophobia. Moreover, MRA imaging is not widely available.

Because it is relatively easy and noninvasive, routine evaluation of coronary arteries should become a standard part of all echocardiographic evaluations, especially during pre-athletic screenings.

Computed Tomography

The initial noninvasive tomographic tool for evaluation of coronary arteries was electron beam computed tomography (EBCT), which was introduced for commercial use in 1984. Since then, there have been rapid advances in cardiac CT technology. Multislice scanners were introduced in 1998.[31] The initial scanners were 4-section multidetector row CTs. Currently, 16-slice and 64-slice scanners are widely available. They provide shorter breath-hold time (25 to 30 seconds for the 16-slice; 6 to 9 seconds for the 64-slice), higher temporal resolution, and thinner slices.[15]

The estimated radiation dose for a coronary angiogram is 3 to 10 mSv, compared with 9 mSv for conventional cardiac CT and 1 to 2 mSv for EBCT.[15] The newer scanners also have the advantage of reducing radiation exposure by lowering the radiation dose during cardiac systole.[32] The next generation of 256-slice scanners holds the promise of faster scanning times, shorter breath-hold times, higher resolution, and radiation doses as low as 2 mSv.

In a comparison of 30 patients with coronary anomalies and 30 controls, EBCT correctly identified all patients with coronary anomalies. Two of the 30 patients with coronary anomalies were misread by coronary angiogram.[18] Similar results have been obtained in other studies.[19] The limitations of EBCT include its limited availability and poor z-axis resolution.[15]

In a study of 17 patients who were referred for a 4-slice or 16-slice cardiac CT because of equivocal findings on a coronary angiogram (16 patients) or an echocardiogram (1 patient), the exact site of the origin and proximal course of the anomalous coronary artery was correctly identified in all patients.[15] Ten patients had an interarterial course of the artery between the aorta and pulmonary trunk. Cardiac CT also accurately differentiated an interarterial course from an intraseptal course, which has important therapeutic implications because the latter is usually benign. The multislice CT was also able to demonstrate angulations or kinking of the vessels at the site of origin. Volume rendering allows 3-dimensional visualization of the epicardial course of the anomalous vessel.[15]

In a more recent evaluation of 16 patients with coronary anomalies

suggested by coronary angiogram, 16-slice or 64-slice coronary CT correctly identified the site of origin and course of the coronary anomalies.[33] Eight patients were scanned using the 64-slice CT. The technique dramatically improved scanning time and temporal resolution of the scans.

Coronary angiography has significant limitations in accurately identifying the proximal course of anomalous coronary arteries and their 3-dimensional relationship with the surrounding structures. Multislice coronary CT will likely emerge as the new gold standard for evaluation of anomalous coronaries, especially with the rapid advances in CT technology.

Management

Various anatomic, physiologic, and patient-dependent variables affect the decision for surgical intervention. For coronary artery abnormalities associated with serious ventricular tachyarrhythmia or myocardial ischemia, surgical intervention is indicated.

Multiple surgical techniques have been utilized, including coronary artery bypass graft (CABG) placement,[34,35] re-implantation of the anomalous coronary to the appropriate sinus,[36,37] and an unroofing procedure (marsupialization) to prevent compression from a change in the angulation.[38,39] Intracoronary stents have also been used as an alternative to CABG in symptomatic patients with ischemia; their structural rigidity may protect the vessel against compression.[40] However, the outcome of intervention in asymptomatic individuals has never been evaluated in a controlled study, and the management of these patients remains controversial. The risk of late coronary insufficiency after coronary repair must be weighed against the risk of sudden death.

All patients with an anomalous coronary that has an interarterial course should have surgical intervention utilizing the unroofing technique wherein the intramural segment is unroofed. The other surgical options are suboptimal, particularly in a teenager or young adult, because of a significant risk of late graft failure with bypass grafting. Patients at high risk for surgical correction should be advised to avoid heavy exercise to prevent complications.

Patients with an intramyocardial course of the anomalous coronary have a fixed and remote alignment of the anomalous coronary as it courses within the muscular sulcus between the great arteries. This condition makes neither unroofing nor re-implantation possible. Surgical intervention in this patient subset should be limited to patients with signs of myocardial ischemia by history or exercise stress testing.

Conclusion

Multislice CT is able to detect the exact origin site and course of an anomalous coronary artery, accurately differentiate an interarterial course from an intraseptal course, show angulations or kinking of the vessels at the site of origin, and provide a 3-dimensional visualization of the epicardial course of the anomalous vessel. This technique will likely emerge as the new gold standard for evaluation of anomalous coronaries, especially with the rapid advances in CT technology. ■

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Main Points

- The most common clinical symptom of coronary artery anomaly is angina or exertional syncope. Physical examination is usually unrevealing in the absence of myocardial infarction or symptoms of ongoing ischemia.
- Approximately 20% of coronary artery anomalies produce sudden death or life-threatening symptoms, including arrhythmias, syncope, and myocardial infarction.
- X-ray coronary angiography has traditionally been considered the gold standard for diagnosing coronary artery anomalies. When x-ray angiography is compared with magnetic resonance angiography or computed tomography, the latter techniques seem better at delineating the origin, proximal course, and relationship with the surrounding structures.
- In a recent evaluation of 16 patients with coronary anomalies suggested by coronary angiography, multislice computed tomography correctly identified the origin and course of the coronary anomalies.
- For coronary artery abnormalities associated with serious ventricular tachyarrhythmia or myocardial ischemia, surgical intervention is indicated.

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