Surgical Therapy for Anomalous Aortic Origin of a Coronary Artery

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Anomalous aortic origin of a coronary artery is a congenital coronary anomaly that carries a risk of a life-threatening cardiovascular event, such as sudden cardiac death or myocardial infarction. Some therapeutic guidelines have been proposed, but specific indications and treatment procedures remain controversial. In general, all patients with anomalous aortic origin of the left coronary artery are indicated for surgical repair, whereas surgical indications for anomalous aortic origin of the right coronary artery have not been established. Various surgical therapies (e.g., coronary artery bypass graft, unroofing, reimplantation, and pulmonary artery translocation) have been reported. The unroofing procedure is presumably a reasonable and safe approach for revascularization in patients with anomalous aortic origin of a coronary artery with a long intramural course, despite the risk of aortic insufficiency or recurrent sudden cardiac arrest. Among surgical procedures, reimplantation is conceivably the most physiologically appropriate and durable procedure. However, the procedure is technically demanding due to the requirement for extensive dissection and vessel mobilization. Currently, optical surgical intervention is proposed based on coronary anatomy. With increasing numbers of treated patients and improved diagnostic tools, based on both anatomy and function, this anomaly may be definitely resolved in the near future.

KEY WORDS: anomalous aortic origin of a coronary artery, coronary artery bypass, reimplantation, sudden cardiac death, unroofing

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Primary prevention of a cardiovascular event is indicated in all patients of anomalous aortic origin of the left coronary artery; however, in patients with anomalous aortic origin of the right coronary artery, surgical revascularization is indicated only in patients with documented coronary ischemia due to an inter-arterial course (class I and level B evidence). When a patient has anomalous aortic origin of the right coronary artery was common among patients in whom sudden cardiac arrest occurred (67%). Furthermore, in seven of 16 (44%) patients with anomalous aortic origin of the right coronary artery patients who experienced sudden cardiac arrest, non-invasive stress tests failed to reveal ischemia in the right coronary artery territory, which was consistent with previous autopsy and individual case reports. Certain subtypes of inter-arterial coronary arteries exhibit malignancy: those with slit-like ostia, which are acutely angled and intramural, and which cross the restrictive substance of the intercoronary pillar. Imaging studies, such as echocardiography, computed tomography angiography, and magnetic resonance imaging, have been unable to reliably delineate these subtleties. In addition, functional studies detecting ischemia, including exercise testing and stress nuclear perfusion imaging, are often nonspecific and unreliable. In such situations, objective surgical indications are difficult to establish. Mainwaring and colleagues proposed that intramural anatomy is the key risk factor associated with ischemic symptoms and suggested that all intramural coronary arteries should undergo surgical repair. In addition, Nagashima and colleagues reported that ages40, male sex, participation in sporting activity, absence of prodromal symptoms, acutely angled (≤30°) take-off from the aorta, and absence of luminal narrowing of the inter-arterial course segment were associated with sudden cardiac arrest in patients in AAOCA with inter-arterial course. Take-off angle of the AAOCA from the ascending aorta might be a key factor involved in sudden death (Fig. 1). On axial acquisition images, where the maximum AAOCA ostium diameter can be observed, an ostium line, defined as an imaginary line representing the ostial diameter, is drawn. From the mid-point of the line, a second line, 5-mm long, is drawn along the center of the AAOCA. The take-off angle of the AAOCA is measured as the angle between these two lines at the mid-point of the ostium line. Eventually, in patients with anomalous aortic origin of the right coronary artery between the aorta and pulmonary trunk, if objective ischemic findings are present, surgical intervention is indicated. If no objective ischemic findings are present, surgical therapy should be carefully considered. Among the reviewed factors, the presence of an intramural segment, a slit-like ostium, and the take-off angle may be useful for determining the indications of surgical intervention.

IV. Technique

1. Coronary artery bypass graft (CABG)

CABG is applicable for all patients with AAOCA. The procedure is simple, but is associated with potential problems including early graft failure and long-term loss of patency. Because most patients who present for surgical correction are young, it has been proposed that bypass with an internal thoracic artery increases the potential for long-term patency, compared with placement of a saphenous vein graft. Unfortunately, without
proximal ligation, the arterial graft often fails because of competitive flow\(^3\). Ligation of the native coronary artery is of significant concern because the initial flow from an internal thoracic artery graft might be insufficient to compensate for acute ligation of a patent vessel, thereby causing increased incidences of hypoperfusion syndrome, ischemia, and mortality\(^1\). Therefore, the CABG may be a less favorable approach based on the above reports\(^4\).

2. Unroofing
When the coronary path is initially within the aortic wall (intramural), surgical unroofing may be feasible\(^17\). First introduced by Mustafa and colleagues\(^18\) in 1981, this procedure moves the anomalous vessel to a more common take-off point and appears to have promising short-term success\(^2\),\(^19\),\(^20\). In this procedure, the affected aortic valve commissure in the unroofing area requires resuspension if intramural segment lies under the commissure; aortic insufficiency can occur after surgery, with an interval of varying length\(^21\),\(^22\). In addition, subclinical changes suggestive of ischemia have been reported to occur after use of the unroofing technique, despite patent neo-coronary ostia, particularly in patients with anomalous aortic origin of the right coronary artery\(^23\). However, the mechanism of ischemia had not been elucidated. Patients with a “short intramural course” have historically undergone a “short unroofing,” and Mery and colleagues described a patient who experienced recurrent sudden cardiac death in this setting\(^24\). The unroofing procedure must move the coronary ostia well beyond the interarterial portion into the correct sinus of Valsalva\(^25\). If there is a short intramural segment, unroofing may increase the size of the ostium, while the vessel remains associated with the incorrect sinus and with a course behind the intra-arterial portion, where it may be compressed\(^26\). In this situation, the coronary artery should be reimplanted within the correct sinus.

3. Reimplantation
Reimplantation avoids the risks associated with CABG and traditional unroofing procedures, but carries its own inherent risks; these include stretching and kinking of the artery, as well as anastomotic stenosis and the long-term effect of a circumferential anastomosis. There have been a few reports of reimplantation in patients for AAOCA\(^12\),\(^25\)-\(^28\). Law and colleagues\(^25\) reported surgical results and outcome after reimplantation for 16 patients with symptomatic anomalous aortic origin of the right coronary artery. In their study, reimplantation was successful in 15 patients (94%) without operative mortality and 10 patients (67%) underwent postoperative computed tomography coronary angiography; none of the patients had stenosis, kinking, or compression of the right coronary artery by the pulmonary artery. Law and colleagues thus concluded that reimplantation technique is safe for treatment of anomalous aortic origin of the right coronary artery and has excellent medium to long-term results regarding symptom-free survival. In contrast to unroofing, the reimplantation technique restores normal anatomy and addresses all the high-risk features of AAOCA, including intramurality, inter-arterial course, and the slit-like ostium\(^25\). Therefore, reimplantation is a promising procedure; however, it is technically demanding because of the extensive dissection and mobilization of vessels required\(^25\). As described above, the reimplantation procedure should be indicated for patients with a short intramural segment, whereas the unroofing procedure should not\(^25\).

Previously, we reported a case involving direct coronary reimplantation of a right coronary artery with the anomalous origin from the sinus of Valsalva in a 41-year-old man with angina (Fig. 2 and 3)\(^29\). After 17 years, the patient remains healthy without any symptoms related to the AAOCA. The reimplanted right cor-

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The anomalous coronary artery has remained patent and the anastomotic shape was excellent in three-dimensional computed tomography at 17-years postoperatively (Fig. 4). Based on the findings during follow-up of our patient, we presume that reimplantation might achieve excellent life-long reconstruction of the coronary artery.

4. Pulmonary artery translocation

A pulmonary artery translocation has been reported to create additional space between the aorta and the pulmonary artery, thus reducing or eliminating compressive and angulation forces on the anomalous coronary artery. The procedure is an alternative option for treating patients with AAOCA, a single coronary ostium, and no intramural component. This procedure can also be used to supplement an unroofing procedure when there remains a possibility of inter-arterial compression.

V. Conclusion and perspectives

The surgical repair of AAOCA is highly effective for relieving the symptoms of myocardial ischemia, if optical surgical repair...
Fig. 5 Proposed algorithm to select optimal surgical intervention for patients with anomalous aortic origin of a coronary artery (AAOCA) requiring intervention based on coronary anatomy. Coronary translocation=Reimplantation (reprinted with permission) 

is adopted and reliably performed for each patient. Currently, optimal surgical intervention is proposed based on coronary anatomy (Fig. 5). With increasing numbers of treated patients and improved diagnostic tools, based on both anatomy and function, as well as multidisciplinary collaborations, this anomaly may be definitively resolved in the near future.

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