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Review article:

Management of anomalous aortic origin of coronary arteries: Review

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Abstract:

Anomalous aortic origin of coronary artery(AAOCA) is a rare congenital cardiac lesion and are generally classified as anomalies of origin, course, supplied region, and size or number of vessels (1,2). AAOCA arise from the aorta by a separate ostium, common ostium, or as a branch vessel of other coronary artey (3). AAOCA are further characterized by 1 of 5 course subtypes as interarterial, subpulmonic (intraconal or intraseptal), pre-pulmonic, retroaortic, or retrocardiac (Central Illustration). Additionally, AAOCA may have an early intramural segment (within the aortic wall), as seen in the majority of interarterial cases.In interarterial course of AAOCA, the anomalous coronary advances between the aorta and pulmonary artery (4). However, a subpulmonic course exits the aorta below the pulmonic valve and traverses the right ventricular outflow tract, pulmonary infundibulum, and interventricular septum.

Keywords: coronary artery disease, aortic origin

Introduction

Anomalous aortic origin of coronary artery(AAOCA) is a rare congenital cardiac lesion and are generally classified as anomalies of origin, course, supplied region, and size or number of vessels (1,2). AAOCA arise from the aorta by a separate ostium, common ostium, or as a branch vessel of other coronary artey (3). AAOCA are further characterized by 1 of 5 course subtypes as interarterial, subpulmonic (intraconal or intraseptal), pre-pulmonic, retroaortic, or retrocardiac (Central Illustration). Additionally, AAOCA may have an early intramural segment (within the aortic wall), as seen in the majority of interarterial cases.

In interarterial course of AAOCA, the anomalous coronary advances between the aorta and pulmonary artery (4). However, a subpulmonic course exits the aorta below the pulmonic valve and traverses the right ventricular outflow tract, pulmonary infundibulum, and interventricular septum (5).

Conventional investigations like invasive angiography and 2D-echocardiography provide limited exposure on the subtypes of course of AAOCA. Among recent advanced radiological studies computed tomography angiography (CTA), magnetic resonance angiography (MRA), and intravascular ultrasound (IVUS) are better in delineating of AAOCA vessels. Although evidence demonstrates that interarterial ALCA and ARCA may be associated with an increased risk of SCD among AAOCA subtypes, the prevalence of AAOCA and their associated absolute risk of

SCD in the general population is unknown. Thus, controversy remains regarding the optimal approach to risk stratify and manage these patients.

EVALUATION OF AAOCA

1. TRANSTHORACIC (TTE) AND TRANSESOPHAGEAL ECHOCARDIOGRAPHY (TEE).

TTE is a commonly used first line of investigation to evaluate patients with suspected or known structural cardiac disease, as it is a noninvasive, rapid, and widely available and cost effective test. Yet TTE has limited accuracy to detect AAOCA, as it is operator dependent and it requires experienced operators to identify coronary ostia. TTE also depends on patient's body built for good image quality. Lastly, TTE also has limited spatial resolution and lacks detailed characterization of AAOCA features and surrounding structures. Transesophageal echocardiography has been used to identify AAOCA and may be useful to visualize abnormal coronaries perioperatively. With the advent of 3-dimensional transesophageal echocardiography, visualization and characterization of AAOCA and their relation to surrounding anatomy may be delineated with slight improvement (6). However TEE is not used routinely for the diagnosis.

2. COMPUTED TOMOGRAPHY CORONARY ANGIOGRAPHY (CTA)/ MAGNETIC RESONANCE ANGIOGRAPHY (MRA).

Currently, coronary CTA and MRA are the only Class I–indicated diagnostic modalities utilized to image AAOCA (7). The choice between these techniques depends on multiple factors, including local expertise, availability and the strengths and limitations of these two modalities. In many centers, CTA is preferred to image AAOCA due to rapid scan times, high spatial resolution, and lower cost in comparison to MRA. CTA has revealed a high diagnostic accuracy to detect coronary artery stenosis when compared with invasive coronary angiogram (8) and has the ability to characterize various other AAOCA features.

Disadvantages of CTA include the need for iodinated contrast agent and hazards of radiation exposure but recent advancements in imaging continue to improve patient safety (9).

3. INVASIVE CORONARY ANGIOGRAM AND INTRA VASCULAR ULTRASONOGRAPHY

The initial course of anomalous coronary arteries is generally not well discernable in majoprity of patients by ICA. Even when ICA identifies AAOCA, many patients are still referred for CTA or MRA to improve visualization. Recent use of specialized ICA catheters which have incorporated IVUS help improve the detection and characterization of AAOCA. As a technique with high spatial and temporal resolution, IVUS offers excellent dynamic imaging. Majority of coronary artery perfusion in left-sided epicardial vessels occurs during diastole, hence stenosis grading is typically measured at the point of maximal narrowing during diastolic phase imaging. Systolic compression of proximal AAOCA vessels may be observed in cases with an early intramural course. As in patients with deep myocardial bridging, prolonged pressure on coronary arteries during systole and early diastole may decrease coronary blood flow. In these cases, IVUS offers superior resolution to image coronary arteries throughout the cardiac cycle. Although IVUS is low risk, engaging AAOCA vessels may be difficult in cases with ostial narrowing, an ostial ridge, or an acute angle takeoff. Additional care is needed during IVUS to distinguish vessel spasm from true narrowing.

4. NONINVASIVE FUNCTIONAL TESTING.

Functional significance of AAOCA can be evaluated with exercise treadmill testing and stress myocardial perfusion imaging. Exercise stress is preferable for evaluating patients with AAOCA when considering that a majority of SCD cases attributed to AAOCA occur with strenuous exercise. Nonetheless, both exercise treadmill testing and stress myocardial perfusion imaging may yield false-positive and false-negative results. Mere absence of ischemia during stress testing cannot be seen as reassuring particularly when potentially high-risk anatomic features are present.

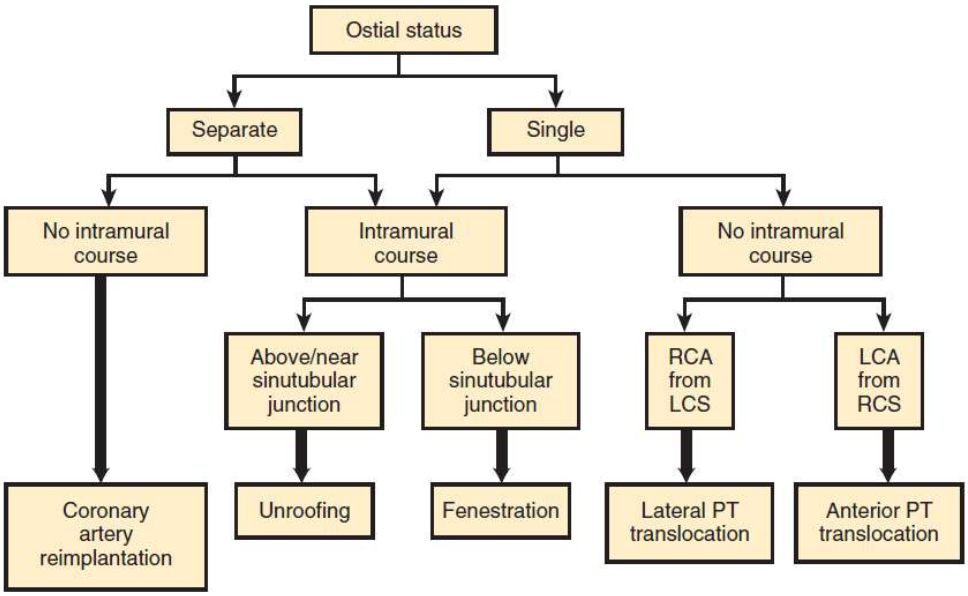
MANAGEMENT

1. SURGICAL REPAIR.

Indications for surgical intervention for isolated AAOCA have evolved but remain controversial [10]. It is recommended to undertake surgical intervention in (a) AAOCA of the LMCA regardless of symptoms, (b) symptomatic patients with AAOCA of the RCA, (c) asymptomatic patients with AAOCA of the RCA with "high-risk" anatomic features (high origin from the aorta, slit-like ostium, acute angled takeoff, and intramural course), (d) AAOCA associated with other surgically correctable cardiac defect.

Mostly non-dominant AAOCA of the RCA is managed non-operatively. Medical optimization for such patients includes exercise restriction along with beta-blockade [11].

Management of the coronary anomalies can be depicted as shown below [12]



Coronary artery bypass grafting (CABG) had been the mainstay of surgical intervention for AAOCA. But in many series, CABG for AAOCA has given disappointing results given the predilection for early graft occlusion in the setting of competitive flow in non-diseased coronary artery [13]. Also CABG along with ligation of the proximal coronary artery to prevent competitive flow is an unattractive option in the pediatric age group.

Coronary artery unroofing of the intramural segment, first described by Mustafa and colleagues [14], has become the surgical treatment of choice for AAOCA. Some surgeons also advocate for routine patch augmentation angioplasty to avoid the risk of residual stenosis of the distal intramural segment (15).

Reconstruction of the neo-ostium and coronary button translocation onto the proper aortic sinus can then be performed when there is separate coronary ostial origin of the malignant artery. A widely patent neo-ostium should then be confirmed by direct inspection and careful passage of a large coronary probe, however this technique is surgically challenging and long mobilization of the coronary artery can lead to injury and kinking of the coronary artery sometimes requiring redo anastomosis or re-exploration.

Lateral translocation of the pulmonary artery onto the left pulmonary artery was described by Rodefeld et al [16] has become the recommended technique when both the coronary arteries share a common ostium or are near to each other not amenable for coronary translocation. His procedure generates additional space between the great arteries and leaves the coronary circulation undisturbed thus decreasing or negating compressive and angulation forces on the anomalous coronary artery. In all cases, care must be taken to avoid iatrogenic injury to the aortic valve commissure and its support.

2. PERCUTANEOUS CORONARY INTERVENTION.

Limited literature is published regarding the use of PCI in patients with interarterial ALCA or ARCA. In a study of 42 predominantly adult patients (mean age 48 years, range 12 to 73 years) with interarterial ARCA undergoing PCI, the rate of in-stent restenosis was 13% by serial IVUS (17). In that study, 29% of patients had recurrent symptoms during a median follow-up period of 5 years.

Although coronary artery bypass graft guidelines state that PCI has been employed as treatment modality in adults with anomalous coronary arteries (18), a recent review by American College of Cardiology/American Heart Association have however recommended surgical procedures as the only modality available for correcting these type of anomalies" (19). Hence, PCI is currently not considered a routine option for revascularization in these patients.

3. EXERCISE RESTRICTION

Recommendations for exercise and discontinuation of competitive sports were highlighted by American College of Cardiology/American Heart Association in 2015.In ARCA patients with symptoms of chest pain or syncope, arrhythmias on ECG, or evidence of ischemia on exercise testing, restriction from all competitive sports is advised while awaiting surgical repair. Although in patients with ARCA without any symptoms or a positive exercise stress test can be advised to continue competitive sports after counselling the patient and relatives regarding the associated risk and benefit. But exceptions may be allowed for participation in light sports. In athletes with ALCA even on absence of symptoms, restriction from all competitive sports is recommended while awaiting surgical repair. After surgery, a return to intense activities may be considered if the patient remains asymptomatic and an exercise stress test shows no evidence of ischemia or cardiac arrhythmias

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