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INVITED EXPERT REVIEW

Management of Adults With Anomalous Aortic Origin of the Coronary Arteries: State-of-the-Art Review

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As a result of increasing adoption of imaging screening, the number of adult patients with a diagnosis of anomalous aortic origin of the coronary arteries (AAOCA) has grown in recent years. Existing guidelines provide a framework for management and treatment, but patients with AAOCA present with a wide range of anomalies and symptoms that make general recommendations of limited applicability. In particular, a large spectrum of interventions can be used for treatment, and there is no consensus on the optimal approach to be used. In this paper, a multidisciplinary group of clinical and interventional cardiologists and cardiac surgeons performed a systematic review and critical evaluation of the available evidence on the interventional treatment of AAOCA in adult patients. Using a structured Delphi process, the group agreed on expert recommendations that are intended to complement existing clinical practice guidelines.

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Because of advances in imaging technologies and the growing use of screening protocols, the number of adult patients with a diagnosis of anomalous aortic origin of the coronary arteries (AAOCA) has grown considerably in recent years.[1] Although current guidelines provide general concepts for diagnosis and treatment, patients with AAOCA present with a wide range of anomalies and symptoms that make general recommendations of limited applicability in clinical practice. In particular, AAOCA can be treated with a large spectrum of interventions, and there is no consensus on the optimal approach to be used.

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Abbreviations and Acronyms

AAOCA = anomalous aortic origin of the coronary arteries
BAV = bicuspid aortic valve
CABG = coronary artery bypass grafting
CMR = cardiac magnetic resonance
CTA = computed tomography angiography
CT-FFR = computed tomography-fractional flow reserve
FFR = fractional flow reserve
IVUS = intravascular ultrasound
L-AAOCA = anomalous aortic origin of the left coronary artery
MI = myocardial infarction
MRA = magnetic resonance angiography
PCI = percutaneous coronary intervention
R-AAOCA = anomalous aortic origin of the right coronary artery
RCA = right coronary artery
SCD = sudden cardiac death

In this paper, a multidisciplinary group of clinical and interventional cardiologists and cardiac surgeons performed a systematic review and critical evaluation of the available evidence on the interventional treatment of AAOCA in adult patients. The group also agreed on expert recommendations by using a structured Delphi process; those recommendations are intended to complement existing clinical practice guidelines.[2,3]

METHODS

Given the lack of individual patients' information, Institutional Review Board approval was not requested for this project.

Details of the systematic review of the literature are reported in the Supplemental Appendix, including Supplemental Table 1. The systematic search identified 3,794 papers. After duplicates were removed, a total of 2,650 records were screened. Of these, 158 publications were selected and informed this document (the Preferred Reporting Items for Systematic Reviews and Meta-Analyses flowchart is shown in Supplemental Figure 1). Because the majority of the evidence derives from case series and case reports, formal grading of the level of evidence was not performed. All supporting data are available from the corresponding author on request.

EXPERT CONSENSUS PROCESS. The consensus process took place between April 24, 2023 and May 1, 2023 following the modified Delphi method (for details on the voting process, see the Supplemental Appendix). As is standard in Delphi documents,[4] the cutoff for agreement was set at 80%.

PANEL SELECTION. A total of 23 experts (16 from the United States, 2 from Italy, 2 from Austria, and 1 each from Canada, Australia, and Switzerland) were invited to participate in the consensus process (Supplemental Table 2), and all agreed to participate. Participants were selected on the basis of clinical and research expertise on AAOCA. Details of the conflicts of interest and relations with industry were collected from each panelist. All panelists participated in the design and revision of the manuscript.

ANATOMY AND PREVALENCE OF AAOCA

CLASSIFICATION. Coronary artery anomalies encompass a wide spectrum of variants, each of them observed in <1% of the general population.[5,6] There is no consensus on anatomical nomenclatures, and the existing classifications often lack details that are very important for risk stratification and clinical decision making.[7,8] The classification proposed by Angelini[9] integrates the features of each anomaly and allows reproducible classification (Supplemental Table 3). Another classification was developed as part of the International Congenital Heart Surgery Nomenclature and Database Project of the European Association for Cardio-Thoracic Surgery and The Society of Thoracic Surgeons and has been incorporated into the International Classification of Diseases-11th revision code.[10]

PREVALENCE. AAOCA can be isolated or observed in syndromic congenital heart disease, such as transposition of the great arteries[11,12] or tetralogy of Fallot (~6% of cases),[13] or in patients with other cardiac anomalies such as bicuspid aortic valve (BAV) (who have a prevalence of AAOCA that is almost double than in patients with tricuspid aortic valve).[14] AAOCA may also occur in patients with syndromic hereditary connective tissue diseases such as Marfan syndrome.[15]

In autopsy series from the general population, the overall prevalence of AAOCA ranges from 0.2% to 0.5%[16,17]; in a national pathology registry of persons who experienced sudden cardiac death (SCD), 30 of 5,100 cases (0.6%) showed coronary anomalies.[18] In selected series of nonathletes who experienced SCD, the prevalence varied from <1%[19-21] up to 17%.[22] In athletes with SCD, the prevalence varies from 5%[23] to 23%.[24-26] In clinical invasive and noninvasive imaging series, the prevalence of isolated coronary artery anomalies ranges from <1% to >5% (Supplemental Table 4).

The most common anomaly is origin of the circumflex artery from the right coronary artery (RCA) or the right sinus of Valsalva; other less common anomalies include origin of the RCA or left coronary artery from another coronary artery, from an inappropriate sinus of Valsalva, or from an ectopic ostium in the ascending aorta.[27] A large study involving cardiac magnetic resonance (CMR) screening of 5,169 asymptomatic adolescents showed a prevalence of 0.44% of AAOCA from the opposite sinus of Valsalva with 0.33% prevalence of AAOCA of the RCA from the left sinus (R-AAOCA) and 0.12% of the left coronary artery from the right sinus (L-AAOCA).[28]

For a summary of the observed prevalence and types of AAOCA arising from the inappropriate sinus of Valsalva, refer to Figure 1.[29]

MECHANISMS OF ISCHEMIA

In patients with AAOCA, mechanisms of myocardial ischemia include both fixed and dynamic stenotic components.[30] Fixed components relate to proximal luminal narrowing and/or ostial abnormalities, whereas dynamic components relate to the course of the coronary artery within the aortic wall or the myocardium and to arterial spasm.

Traditionally, the interarterial course between the aorta and the pulmonary artery was thought to be the main ischemic mechanism, resulting from compression between the 2 vessels.[9] However, the current general thought is that the interarterial course per se is unlikely to cause significant coronary compression (because the pressure is higher in the aorta and in the coronary arteries compared with the pulmonary circulation) and is rather a surrogate for the presence of an intramural segment (within the aortic wall) that is the true mechanism for ischemia (although cases of aborted SCD have been described in patients with an interarterial course and no intramural segment or a very short intramural segment).[31]

Under resting conditions, the intramural, oval vessel portion of the AAOCA shows a phasic lateral compression through the cardiac cycle with pronounced compression in systole.[32] Under strenuous exercise, with augmented aortic wall stress secondary to increased stroke volume, blood pressure, and heart rate, the dynamic compression may lead to hypoperfusion and myocardial ischemia resulting from the reduced cross-sectional area of the oval vessel (compared with a round vessel) and higher resistance to flow (on the basis of the Hagen-Poiseuille law).

As to ostial abnormalities, an important anatomical high-risk feature is the slit-like ostium, which may or may not be associated with an intramural segment.[33] When an acute takeoff angle (<45°) coexists with ostial abnormalities, an intermittent valvelike coronary obstruction can occur.[34,35] This is most pronounced in systole and may become clinically relevant under stress conditions when coronary perfusion also occurs in systole as a result of shortening of the diastolic filling interval. Other potential stenotic components include the course behind the intercommissural pillar of the aortic valve,[34,36] ostial hypoplasia,[37] or, in rare cases, stenosis of the coronary artery distal to the intramural segment.

Coronary spasm is rarely observed in transseptal AAOCA, but it may occur in adults, and myocardial ischemia has been reported in young patients.[38-41]

Ischemia does not occur consistently in the presence of high-risk anatomical features, and in the same patient ischemia does not reproducibly occur with physical activity,[42] findings suggesting that other factors (eg, volume status, type of exercise, and others unknown variables) may play an important trigger role.[43,44]

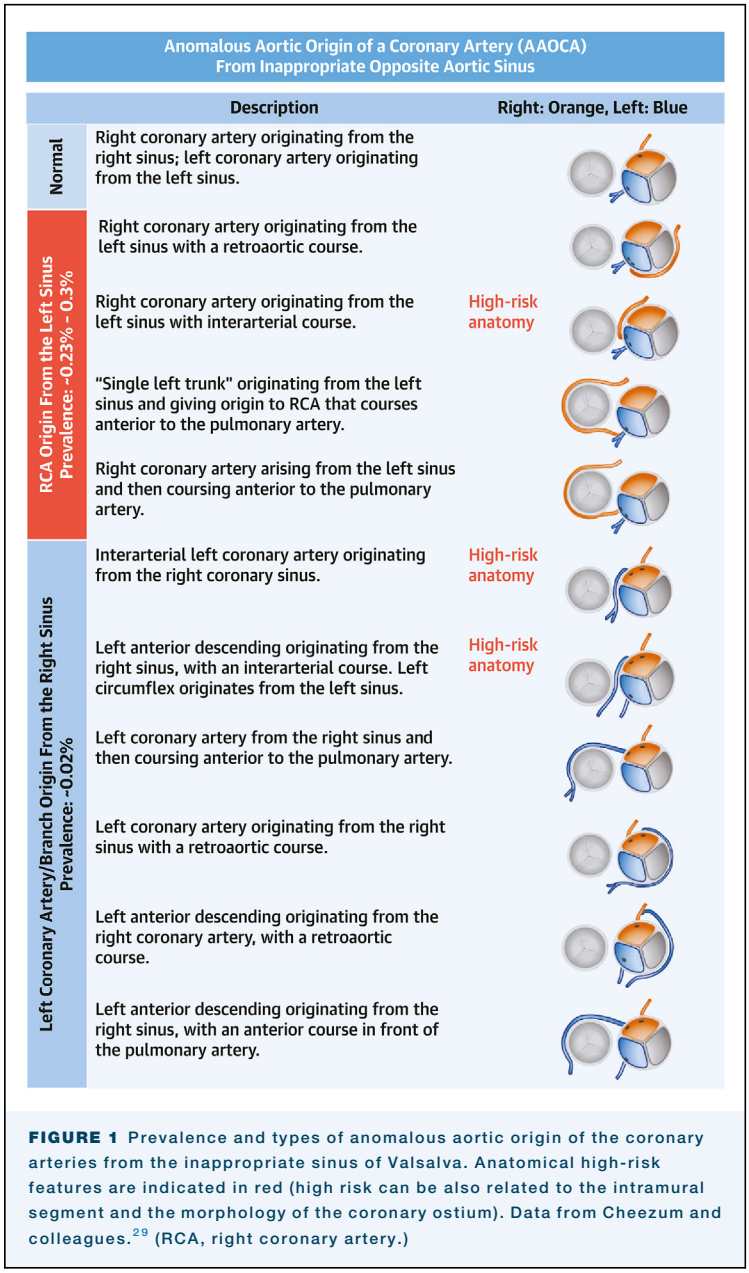


FIGURE 1 Prevalence and types of anomalous aortic origin of the coronary arteries from the inappropriate sinus of Valsalva. Anatomical high-risk features are indicated in red (high risk can be also related to the intramural segment and the morphology of the coronary ostium). Data from Cheezum and colleagues.[29] (RCA, right coronary artery.)

EFFECTS OF AAOCA ON THE OPERATIVE RISK OF PATIENTS UNDERGOING NONCORONARY CARDIAC SURGERY

The perioperative risk associated with cardiac surgery for noncoronary disease in the presence of AAOCA depends on the origin and the course of the anomalous coronary artery. The most frequently described variant in patients undergoing BAV surgery is the anomalous

TABLE 1 Key Features of the Main Imaging Modalities for Work-Up of Anomalous Aortic Origin of the Coronary Arteries

				Nuclear		Invasive Angiography		
	Echo	CTA	CMR	SPECT	PET	Conventional Angiography	Intravascular Ultrasound	Invasive Functional Testing
Imaging factors[a]								
Spatial resolution	+++	++++	++	–	–	+++++	+++++	–
Temporal resolution	++++	+	++	–	–	++++	+++	–
Ionizing radiation exposure	–	++	–	+++	++	++	N/A[b]	N/A[b]
Nephrotoxic contrast exposure	–	+	–	–	–	+	N/A[b]	N/A[b]
Anatomical delineation								
Anomalous coronary origin	++	+++++	++++	–	–	+++	++++	–
Takeoff angle	++	+++++	+++	–	–	+++	+++	–
Ostial narrowing	+	++++	++	–	–	++	+++++	–
Intramural segment	+	++++	++	–	–	+	++++	–
Proximal narrowing and elliptical shape	+	++++	++	–	–	++	++++	–
Assessment of concomitant CAD[c]	–	+++++	++++	+++	++++	+++++	N/A[b]	N/A[b]
Tissue characteristics								
Ischemia[d]	+++	–	++++	+++	++++	–	–	++++
Infarction	+	+	++++	++	+++	–	–	–

[a]Grading on the basis of systematic review of published studies in AAOCA (which lacks randomized prospective comparator data), as well as generally established concepts in cardiovascular imaging; A structured 5-point grading scale (+, ++, +++, ++++, +++++) was used and is intended to provide an overview of relative strengths of given modalities; [b]Intravascular ultrasound and invasive functional testing are typically performed in conjunction with conventional angiography and can be useful to provide targeted assessment of AAOCA with minimal increase in radiation or contrast exposure; [c]Coronary CTA and conventional angiography provide comprehensive anatomical assessment of concomitant atherosclerotic coronary disease; nuclear imaging and stress CMR can detect flow-limiting stenosis or evidence of previous infarction in territories distinct from the AAOCA; [d]Robust comparative data for inducible ischemia testing in AAOCA are lacking; thus, this table depicts the ability of each modality to detect flow-limiting ischemia in atherosclerotic coronary disease cohorts, but whether individual diagnostic modalities or individual stress approaches (eg, exercise vs dobutamine stress) have specific advantages in AAOCA is incompletely studied. AAOCA, anomalous aortic origin of the coronary arteries; CAD, coronary artery disease; CMR, cardiacmagnetic resonance; CTA, computed tomography angiography; Echo, echocardiography; N/A, not applicable; PET, positron emission tomography; SPECT, single-photon emission tomography.

circumflex artery arising from the right coronary sinus.[14,45] The vessel usually takes a posterior course to the atrioventricular groove and can be injured at the time of the aortotomy. Inadequate myocardial protection and compression, distortion, and suture impingement of the AAOCA have also been reported.[46] Measures to avoid these complications include downsizing the aortic prosthesis or implanting the valve in a supra-annular position, although this may lead to suboptimal hemodynamics. Sharp dissection and mobilization of the anomalous artery in proximity to the aortic annulus may be technically challenging but allow optimal aortic valve replacement.[47,48] Because the anomalous artery may run in proximity to the mitral valve annulus, there is also a risk of compression when the mitral annulus is heavily calcified[49] and with mitral valve surgery.[50,51] Extrinsic compression of an anomalous circumflex artery in the atrioventricular groove has also been reported with transcatheter aortic valve interventions.[52]

When aortic root replacement is performed in patients with an anomalous coronary ostium very close to the aortic commissure, the ostium can be left attached to the commissure to reduce the risk of distortion and postoperative ischemia,[53] but proximal ligation of the AAOCA and coronary artery bypass grafting (CABG) may be the safest strategy in complex cases.

Variations in size and length of the coronary arteries (not AAOCA) can also increase the operative risk of cardiac surgical procedures. In patients with BAV, the left main coronary artery is often short, and delivery of selective cardioplegia may be difficult. Similarly, a small RCA may lead to inadequate cardioplegia delivery and right ventricular protection.

IMAGING

Risk stratification of AAOCA patients relies on imaging to assess anatomy or evidence of myocardial ischemia. A summary of the key features of the main imaging modalities for AAOCA work-up is reported in Table 1. The main imaging approaches are outlined here.

ANATOMICAL DELINEATION. Invasive angiography. Invasive coronary angiography enables identification of AAOCA origin with high spatial and temporal resolution, as well as assessment of dynamic changes in luminal caliber throughout the cardiac cycle. This approach allows quantification of coronary flow at baseline and during pharmacologic challenges (eg, dobutamine), thereby providing insight into the adequacy of coronary anatomy to accommodate physiologic stressors. As described later, intravascular ultrasound (IVUS) may provide additional information with respect to luminal

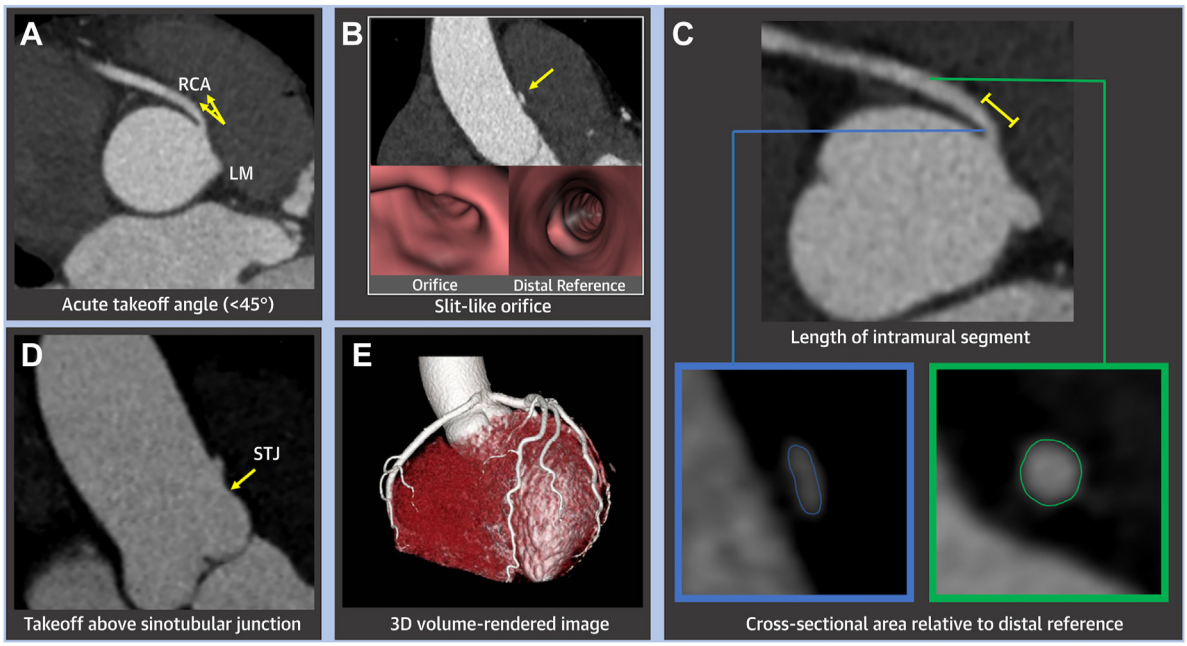


FIGURE 2 Computed tomography scan of anomalous aortic origin of the right coronary artery with an interarterial course and an intramural segment. (A) Multiplanar reformat image showing an acute takeoff angle (<45°) of the anomalous aortic origin of the right coronary artery (RCA) (arrows). (B) Multiplanar reformat image and virtual computed tomography angiography reconstruction images of a RCA with anomalous aortic origin (arrow) revealing a slit-like orifice as compared with the distal reference vessel. (C) Multiplanar reformat and double-oblique short-axis images depicting the intramural segment (yellow line) of the RCA with anomalous aortic origin, with approximately 50% ostial narrowing of the intramural segment relative to the distal reference (outlined areas). (D) Multiplanar reformat image showing that the takeoff level of the RCA with anomalous aortic origin is above the sinotubular junction (STJ) and the left sinus of Valsalva. (E) Three-dimensional (3D) volume-rendered image of the anomalous aortic origin of the right coronary artery and normal left coronary system. (LM, left main coronary artery.)

narrowing and vessel wall characteristics. Disadvantages include its invasiveness (prohibiting use for population-based screening), radiation exposure, and/or technical challenges relating to identification of AAOCA course and dynamic compression and limited information on spatial relationship with the surrounding structures.[54,55]

Noninvasive angiography. Cardiac computed tomography angiography (CTA) has transformed noninvasive screening and evaluation of AAOCA.[56] Widely available cardiac CTA technologies provide submillimeter spatial resolution for delineation of coronary anatomy in relation to the great arteries and myocardial geometry,[1,57,58] to assess risk modifiers such as origin, course, luminal narrowing, and coronary atherosclerosis. Multiple studies have shown cardiac CTA to provide high accuracy for identifying coronary intramural segment and other high-risk features.[59] Regarding risk stratification, registry data reported anatomical factors associated with ischemia to include the presence and length of an intramural segment and a high or slit-like orifice (Figures 2A-2E).[60] However, a large cohort study reported no difference in the presence and length of the intramural segment between young R-AAOCA patients with and without evidence of stress ischemia.[44] Cardiac CTA has also been used to identify concomitant atherosclerosis in AAOCA patients, a finding that is important to inform decision making.[61]

Magnetic resonance angiography (MRA) provides an alternative for noninvasive anatomical delineation of AAOCA.[28,62-64] Key advantages include the lack of ionizing radiation, the lack of a definitive need for contrast material, and adjunctive data attainable within a single examination, including cardiac structure, function, and myocardial tissue characterization. However, MRA can be limited by factors such as a closed-space imaging environment, lower spatial resolution than cardiac CTA, prolonged acquisition time, and susceptibility artifact produced by metallic implants (surgical staples, stents) that can compromise surveillance after interventions. Compared with cardiac CTA, lower spatial resolution often limits precise characterization of high-risk anatomical features. Cardiac CTA and MRA data are typically attained at a single phase of the cardiac cycle and thus provide a limited window into dynamic coronary compression.

Echocardiography. Transthoracic echocardiography use is largely restricted to screening of pediatric populations, given its limited ability to delineate coronary anatomy in adults.[65,66] Additional disadvantages include limited information on slit-like origin and intramyocardial segment as well as coronary dominance.[67]

FUNCTIONAL ASSESSMENT. Current U.S. and European guidelines recommend initial physiologic evaluation in patients with AAOCA, and European guidelines explicitly recommend nonpharmacologic functional imaging (eg, nuclear, echocardiography, or CMR exercise stress) to assess ischemia and inform treatment decisions (Table 2).[2,3] However, no study has prospectively tested the prognostic value of ischemia assessment in patients with AAOCA, and whether intervention mitigates clinical event risk remains unresolved.

As a general principle, exercise (>90% of the predicted heart rate) or dobutamine is preferred over vasodilator stress testing given that the former approaches are believed to better reflect the dynamic hemodynamic conditions resulting in ischemia in patients with AAOCA.[30] However, treadmill testing has been shown to be limited as a tool to stratify risk with AAOCA.[44,68]

The main functional methods for ischemia assessment in AAOCA are outlined here.

Perfusion imaging. Myocardial perfusion imaging can be performed using a variety of imaging modalities, each of which has been investigated in small AAOCA cohorts where ischemia has been tested in relation to anatomical

TABLE 2 Summary of Recommendations for Treatment of Patients With Anomalous Coronary Arteries From the AHA/ACC and ESC Guidelines

Recommendation	Class of Recommendation	Level of Evidence
Diagnostic work-up		
U.S. AHA/ACC 2018 guidelines		
Coronary angiography, using catheterization, CT, or CMR, is recommended for evaluation of anomalous coronary artery.	I	C
Anatomical and physiologic evaluation should be performed in patients with anomalous aortic origin of the left coronary from the right sinus and/or right coronary from the left sinus.	I	C
E.U. ESC 2020 guidelines		
Nonpharmacologic functional imaging (eg, nuclear study, echocardiography, or CMR with physical stress) is recommended in patients with coronary anomalies to confirm/exclude myocardial ischemia.	I	C
Anomalous aortic origin of the coronary artery		
U.S. AHA/ACC 2018 guidelines		
Surgery is recommended for AAOCA from the left sinus or AAOCA from the right sinus for symptoms or diagnostic evidence consistent with coronary ischemia attributable to the anomalous coronary artery.	I	B
Surgery is reasonable for anomalous aortic origin of the left coronary artery from the right sinus in the absence of symptoms or ischemia.	IIa	C
Surgery for AAOCA is reasonable in the setting of ventricular arrhythmias.	IIa	C
Surgery or continued observation may be reasonable for asymptomatic patients with an anomalous left coronary artery arising from the right sinus or right coronary artery arising from the left sinus without ischemia or anatomical or physiologic evaluation suggesting potential for compromise of coronary perfusion (eg, intramural course, fish mouth–shaped orifice, acute angle).	IIb	B
E.U. ESC 2020 guidelines		
Surgery is recommended for AAOCA in patients with typical angina symptoms who present with evidence of stress-induced myocardial ischemia in a matching territory or high-risk anatomy.[a]	I	C
Surgery should be considered in asymptomatic patients with AAOCA (right or left) and evidence of myocardial ischemia.	IIa	C
Surgery should be considered in asymptomatic patients with L-AAOCA and no evidence of myocardial ischemia but high-risk anatomy.[a]	IIa	C
Surgery may be considered for symptomatic patients with AAOCA even if there is no evidence of myocardial ischemia or high-risk anatomy.[a]	IIb	C
Surgery may be considered for asymptomatic patients with L-AAOCA without myocardial ischemia and without high-risk anatomy[a] when they present at young age (<35 years).	IIb	C
Surgery is not recommended for R-AAOCA in asymptomatic patients without myocardial ischemia and without high-risk anatomy.[a]	III	C

[a]High-risk anatomy includes features such as an intramural course and orifice anomalies (slit-like orifice, acute-angle takeoff, orifice >1 cm above the sinotubular junction). AAOCA, anomalous aortic origin of the coronary arteries; AHA/ACC, American Heart Association/American College of Cardiology; CMR, cardiovascular magnetic resonance; CT, computed tomography; ESC, European Society of Cardiology; E.U., European Union; L-AAOCA, anomalous aortic origin of the left coronary artery; R-AAOCA, anomalous aortic origin of the right coronary artery; U.S., United States. Data from Stout and colleagues[2] and Baumgartner and colleagues.[3]

risk surrogates. Although CMR studies have shown an association between high-risk anatomical variations and ischemia,[69,70] studies using single-photon emission computed tomography perfusion imaging have reported conflicting results.[71,72] Studies using positron emission tomography have shown perfusion abnormalities to be frequently associated with causes other than anomalous coronary anatomy.[73] Exercise stress protocols for both positron emission tomography[74] and CMR[75] may hold promise for AAOCA evaluation in experienced centers. Importantly, current perfusion technologies can assess ischemia only when the myocardial territory served by the AAOCA is within the left ventricle, but they are limited for assessment of right ventricular perfusion.

IVUS and invasive functional imaging. Methods for invasive coronary assessment using intravascular imaging (primarily IVUS) and invasive functional testing can provide novel insights into mechanisms of ischemia and offer adjunctive opportunities for risk stratification of patients with AAOCA. IVUS can assess the presence and length of the intramural segment, provides a detailed and dynamic assessment of coronary cross-sectional area at high spatial and temporal resolution, and can be paired with pharmacologic stress testing to quantify changes in cross-sectional area with simulated exercise.[76]

Invasive angiography can also be paired with functional testing to assess for coronary flow impairment secondary to AAOCA[40]; reasonable agreement has been reported between stress CMR and fractional flow reserve (FFR).[77] However, current cutoff values for intracoronary flow derive largely from studies in adults with atherosclerotic heart disease and may not be applicable to patients with AAOCA.

Diffuse coronary artery spasm has been shown to occur in some patients with AAOCA[38] and may be a causative factor for ischemia. Acetylcholine provocative testing may be considered to inform treatment decisions.

CMR tissue characterization. Evidence of myocardial infarction (MI) can be used to stratify risk in patients with AAOCA because autopsy data have shown MI to be present in many AAOCA patients with SCD.[18,68] In small studies using CMR, the prevalence of MI has been reported to be less than in autopsy series.[78]

Computed tomography fractional flow reserve. Limited data on the utility of computed tomography FFR (CT-FFR) (Figures 3A and 3B) for AAOCA are available. In an initial exploratory study, CT-FFR<0.80 was associated with surface narrowing, vessel eccentricity, and decreased angulation, and CT-FFR yielded good performance for identifying AAOCA with an intramural segment.[79] However, nearly one-half of cardiac CTA data sets were deemed inadequate for CT-FFR, thus raising uncertainty with respect to its generalizability. Moreover, CT-FFR protocols have been validated in patients with coronary artery disease and may be inadequate for patients with AAOCA.[80]

Computational modeling. Novel insights into anatomical and functional aspects of AAOCA can potentially be gained through reconstructions such as 3-dimensional representations and adjunctive computational modeling.[81] A small comparative study found 3-dimensional reconstructions to yield incremental accuracy to both cardiac CTA and echocardiography for assessment of coronary anatomy determined intraoperatively (Figures 4A-4F).[66,82] Computational methods such as fluid structure interaction modeling allow integration of multidimensional geometry with coronary flow physiology. Such techniques hold promise for replication of preoperative flow dynamics and relief of abnormal flow patterns after intervention in AAOCA, but further studies are warranted.[83,84]

CLINICAL DECISION MAKING

In patients with AAOCA, current U.S. and European guidelines recommend intervention in the presence of symptoms or evidence of ischemia. Guidelines differ on the emphasis placed on high-risk anatomical features as indications for intervention. U.S. guidelines uniformly recommend surgery for patients with L-AAOCA and for patients with R-AAOCA with associated symptoms, evidence of ischemia, or arrhythmias without specific mention of how anatomical features modify the strength of the indication. European guidelines provide the strongest recommendation for intervention in patients with typical anginal symptoms and evidence of ischemia (in territories subtended by the anomalous coronary and/or high-risk anatomical findings) (Table 2).[2,3] However, an array of patient-specific factors should be considered to tailor risk-benefit assessment and optimize clinical decision making (Figure 5).

Shared decision making, taking into account available evidence, individual characteristics, and patient preference, is of paramount importance.

ANATOMICAL FEATURES. Anatomical features of AAOCA are currently important in clinical decision making, although our understanding of the interaction between these features and SCD risk remains incomplete.

Origin. Autopsy data show that although the population prevalence of R-AAOCA exceeds that of L-AAOCA on the order of 5- to 10-fold, the prevalence of L-AAOCA in young athletes with SCD exceeds that of R-AAOCA by approximately 5 times.[68] This finding is reflected in current guidelines, where the indication to treatment is stronger for L-AAOCA (Table 2).[2,3] The indications for intervention of R-AAOCA in the absence of symptoms

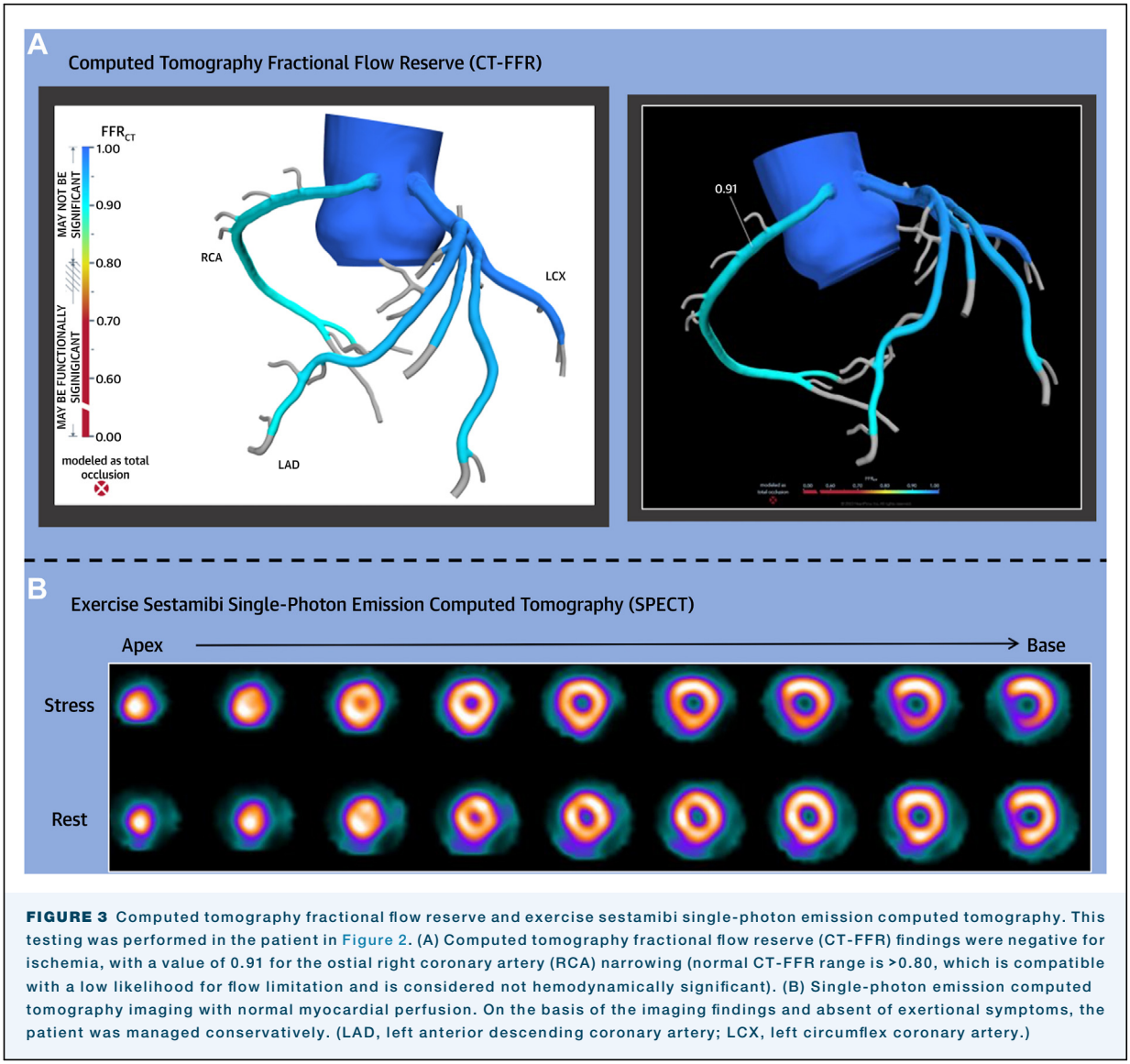


FIGURE 3 Computed tomography fractional flow reserve and exercise sestamibi single-photon emission computed tomography. This testing was performed in the patient in Figure 2. (A) Computed tomography fractional flow reserve (CT-FFR) findings were negative for ischemia, with a value of 0.91 for the ostial right coronary artery (RCA) narrowing (normal CT-FFR range is >0.80, which is compatible with a low likelihood for flow limitation and is considered not hemodynamically significant). (B) Single-photon emission computed tomography imaging with normal myocardial perfusion. On the basis of the imaging findings and absent of exertional symptoms, the patient was managed conservatively. (LAD, left anterior descending coronary artery; LCX, left circumflex coronary artery.)

or positive functional testing results is controversial, given the relatively low relative risk of SCD (although likely higher than the anatomically normal population).

Course and other high-risk anatomical features. Data suggest that the course of the AAOCA modifies the risk of SCD. Although at the population level retroaortic (0.28%) and interarterial R-AAOCA (0.23%) courses are relatively prevalent when compared with interarterial L-AAOCA (0.03%),[29] they are not as well represented in autopsy series of SCD, and they are less frequently associated with symptoms.[26,85,86] Among the different anomalous AAOCA courses (ie, retroaortic, prepulmonic, interarterial, transseptal, retrocardiac), the interarterial course (and in particular interarterial L-AAOCA) dominates autopsy series of SCD. Although reports of symptoms related to myocardial bridge physiology or spasm and myocardial ischemia in patients with transseptal L-AAOCA have been reported, the risk associated with this anomaly remains unclear.[38,39,87]

Further risk stratification can be based on the presence or absence of high-risk anatomical features, such as the presence and length of the intramural segment, a slit-like ostium, proximal narrowing, an acute takeoff aortic angle, and a thickened intercoronary pillar. All these features have been associated with symptoms,[88,89] ischemia on functional testing,[90] and SCD in autopsy series,[34,86,91] and they must be considered in treatment decisions.

In summary, although some types of AAOCA with high-risk anatomical features are likely hemodynamically relevant, have been linked to myocardial ischemia and/or SCD, and probably benefit from

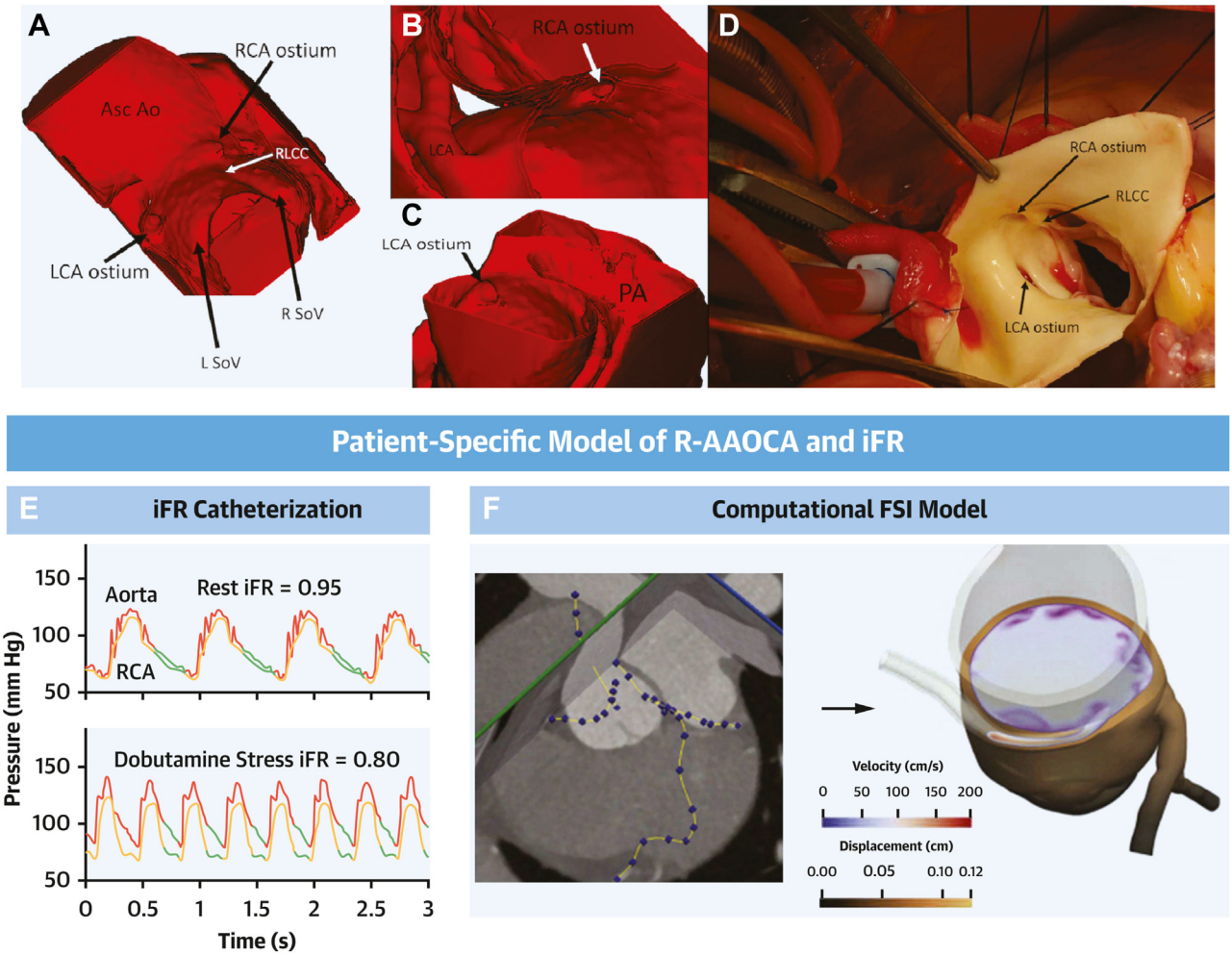


FIGURE 4 A 3-dimensional model and intraoperative findings on a representative patient with anomalous aortic origin of the right coronary artery. (A-C) The 3-dimensional model accurately represents the anomalous coronary position as well as ostial characteristics that correlated well with the (D) intraoperative findings. (E) A patient-specific model of anomalous aortic origin of the right coronary artery (R-AAOCA) shows the instant wave-free ratio (iFR) hemodynamic findings, and (F) shows computational modeling. (Asc Ao, ascending aorta; FSI, fluid structure interaction; LCA, left coronary artery; L SoV, left sinus of Valsalva; PA, pulmonary artery; RCA, right coronary artery; RLCC, right left coronary commissure; R SoV, right sinus of Valsalva.) Modified with permission from Farooqi and colleagues[66] and Jiang and colleagues.[82]

interventions, there are other types of AAOCA that are generally considered benign, unlikely to induce myocardial ischemia, and these patients should not be referred for treatment (among them patients with AAOCA with a prepulmonic or retroaortic course and AAOCA with high takeoff without high-risk features). Similarly, patients with interarterial R-AAOCA are unlikely to benefit from intervention in the absence of anatomical high-risk features, symptoms, or evidence of ischemia.

SYMPTOMS. In adult patients with AAOCA, multiple causes of angina-like syndromes may coexist (coronary atherosclerosis, microvascular coronary disease, noncardiac chest pain),[73] and symptoms related to AAOCA may be atypical, so the role of symptoms in informing treatment decisions is unclear.[68]

Evidence of stress-induced ischemia or MI in the territory subtended by the AAOCA in a patient with typical anginal symptoms provides the strongest level of evidence supporting treatment. The inability to evoke ischemia on physiologic testing, atypical symptoms, and the presence of risk factors for coronary vascular disease decrease the strength of this support. Nonsustained ventricular tachycardia is a Class IIb indication for intervention in the U.S. guidelines.[2]

A further important caveat is that many patients who experience SCD have no clinically reported symptoms before their event, thus indicating that the absence of

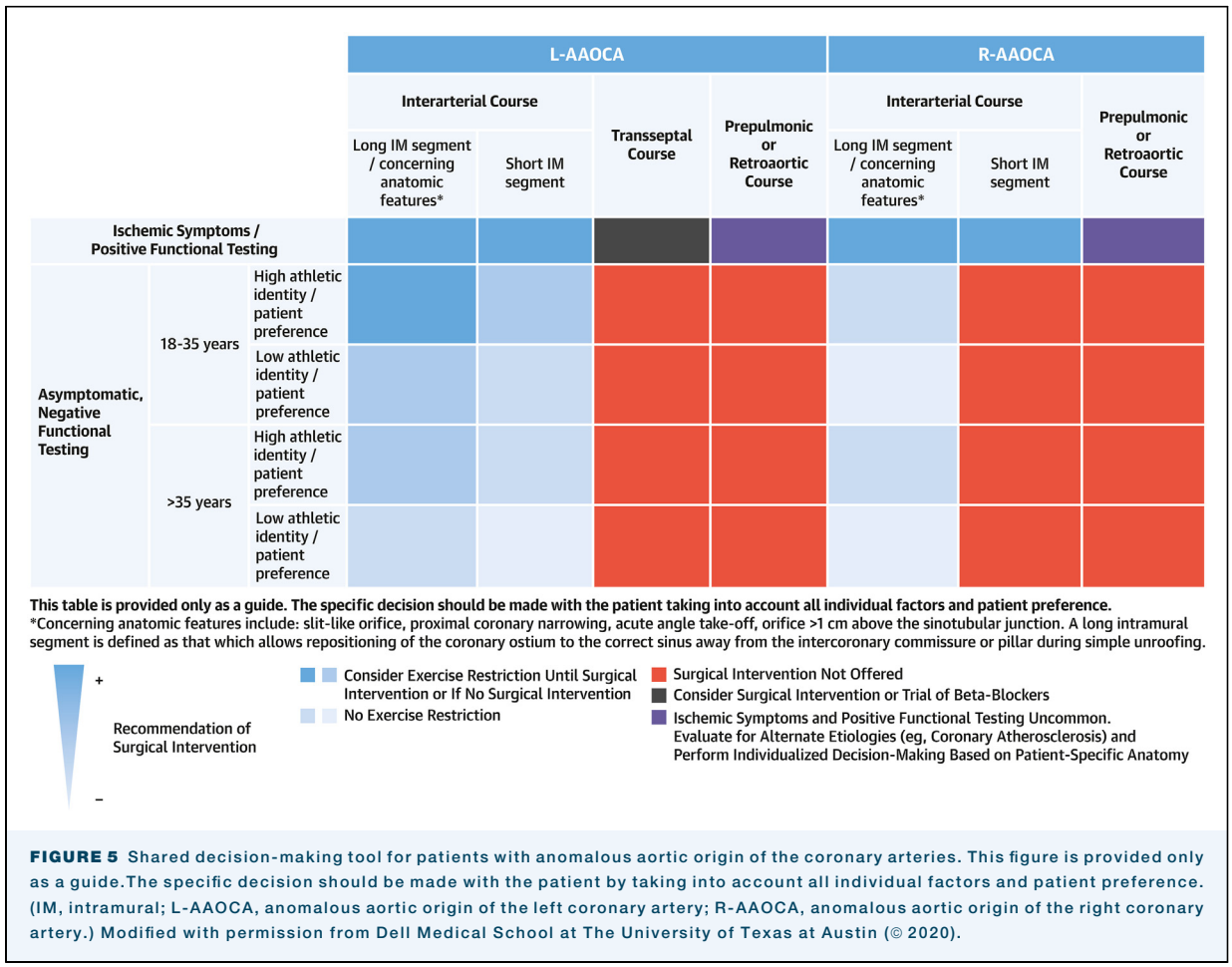


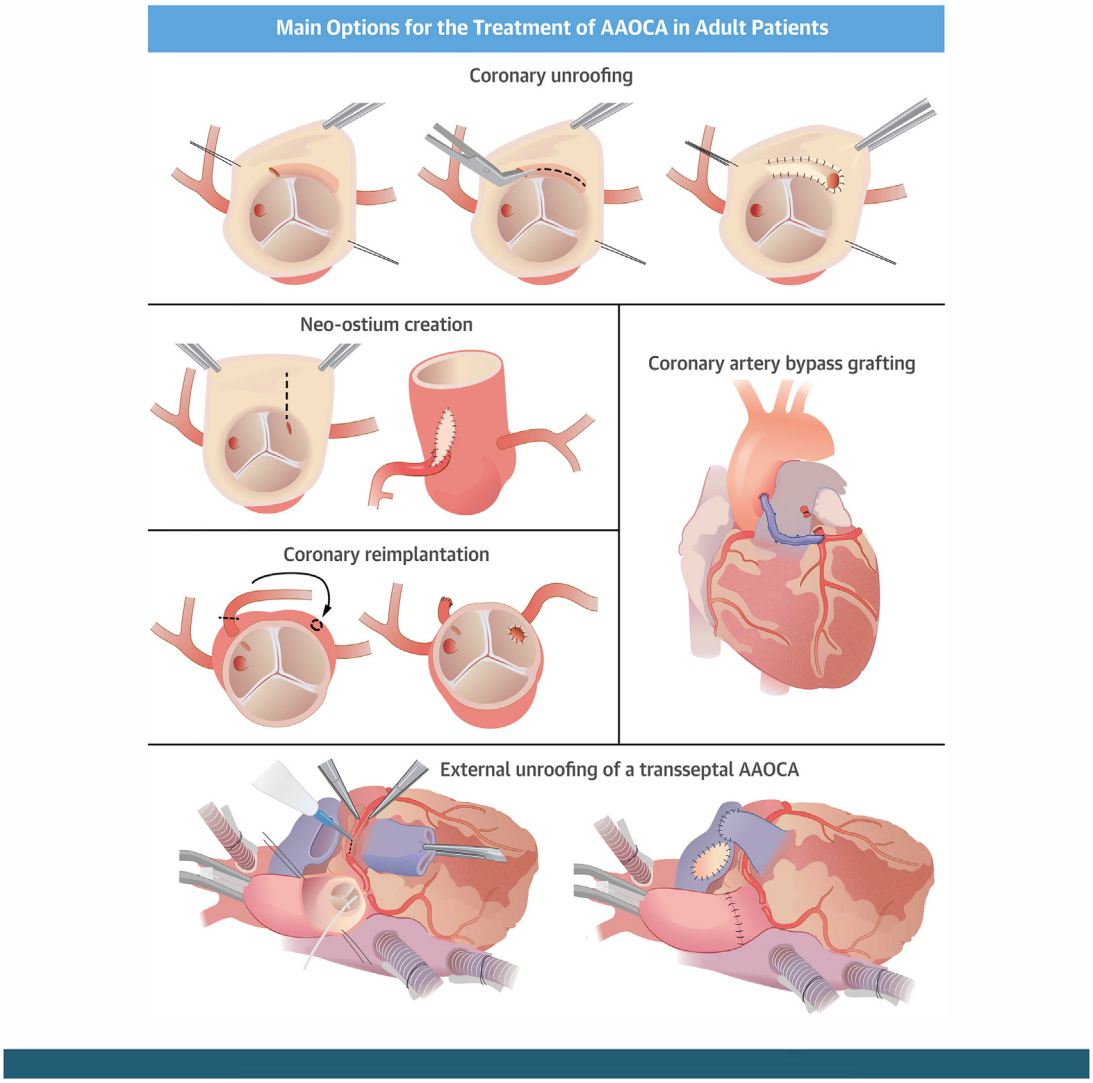
FIGURE 5 Shared decision-making tool for patients with anomalous aortic origin of the coronary arteries. This figure is provided only as a guide.The specific decision should be made with the patient by taking into account all individual factors and patient preference. (IM, intramural; L-AAOCA, anomalous aortic origin of the left coronary artery; R-AAOCA, anomalous aortic origin of the right coronary artery.) Modified with permission from Dell Medical School at The University of Texas at Austin (© 2020).

symptoms is insufficient to reassure against acute events.[29,68,86] Conversely, middle-aged and older adult patients with newly detected AAOCA have a low risk of adverse outcomes with conservative treatment,[92] and it is likely that most asymptomatic patients with AAOCA (in particular R-AAOCA with no signs of ischemia) can be managed with a conservative wait-and see approach on the basis of a multidisciplinary evaluation including the patient.

AGE AND ACTIVITY LEVEL. Age and activity level are important modifiers of SCD risk. Many of the available data are derived from autopsy series of young competitive athletes, and most cases of SCD have been described in patients <30 years of age.[93] In a study of middle-aged patients (mean age 56 ± 11 years) who were undergoing clinically indicated cardiac CTA, the presence of AAOCA was not associated with an increased risk of major adverse cardiac events compared with matched control subjects.[92] However, SCD has also been reported in older patients,[18] and older age alone does not exclude the need for intervention. European guidelines modify the strength of recommendation for intervention after 35 years of age,[3] although in the absence of clear supportive data.

It is reasonable to restrict activity of patients with newly diagnosed AAOCA and high-risk features during work-up, and the desire to return to high-level activity in the face of uncertain risk may justify the decision to proceed with intervention in equivocal cases. Although most data on SCD are derived from studies in competitive athletes, it should also be acknowledged that SCD attributed to AAOCA does not exclusively occur during or immediately following exertion.[18,93] Cases of spontaneous dissection of AAOCA have also been reported.[94-96]

SURGICAL RISK. Surgical risks should be incorporated in decision making, by acknowledging that in the hands of experienced operators, the risks of major procedural complications and death are extremely low, although risks vary with the surgical approach.[97] In older adults with frailty and multiple comorbidities, surgical risk may be augmented, and the benefits of repair are



VISUAL ABSTRACT Main surgical techniques for treatment of anomalous aortic origin of the coronary arteries in adult patients. Coronary unroofing, neo-ostium creation, coronary artery bypass grafting, coronary reimplantation, and external unroofing of a transseptal coronary artery with anomalous aortic origin. (AAOCA, anomalous aortic origin of the coronary arteries.)

likely blunted by decreased activity level, shorter life expectancy, and competing risk factors.

PATIENT PREFERENCE. Quality of life is negatively affected by clinical symptoms, and even in absence of physical symptoms, AAOCA patients may experience substantial mental stress from knowledge of a condition that has been associated with SCD.[98] A patient's desire to avoid anxieties related to SCD risk and exercise restriction can influence treatment decisions. In competitive athletes, the therapeutic approach to AAOCA may also have significant implications for the athlete's career and general well-being.

SURGICAL TECHNIQUES

A simplified image of the most common surgical techniques used to treat AAOCA is provided in the Visual Abstract.

UNROOFING. Unroofing is the most commonly used technique to repair AAOCA and can be applied in cases with an intramural component (although the exact determination of the length of the intramural segment can be challenging on the basis of preoperative imaging).

In most cases, the intramural segment runs superior to the left or right aortic commissure, and the repair can be performed without interference with the aortic valve

through incision and marsupialization of the endothelial tissue flap and limited resection when needed. The incision must extend for the entire length of the intramural segment and is usually between 5 and 15 mm in length.

In some patients, the intramural tunnel runs below the aortic valve commissure or across it. In those cases, it is possible to create a neo-ostium on the other side of the commissure (partial unroofing into the correct sinus). A more complex approach is to detach the aortic valve commissure, complete the unroofing, and then resuspend the commissure, but this technique carries the risk of aortic insufficiency resulting from valve damage or imperfect commissural resuspension.[99]

The results of unroofing are excellent, with no reported operative mortality.[100,101] Potential complications are aortic dissection (from insufficient marsupialization or trauma), aortic regurgitation from commissure prolapse, and recurrent stenosis, which is generally related to incomplete unroofing. There have been reports of residual compression and ischemia after adequate unroofing of a short intramural segment when the artery runs behind or close to the intercoronary pillar.[102]

OSTIOPLASTY. The goal of ostioplasty is to create an adequately sized ostium within the appropriate sinus of Valsalva; the procedure is often combined with unroofing. Ostioplasty is indicated in patients with a slit-like ostium or a tight exit angle of the AAOCA from the aorta. At surgery, the ascending aorta is incised above the exit of the AAOCA, and the incision is extended vertically into the coronary artery.[103] A triangular patch of autologous pericardium is used to enlarge the coronary ostium and eliminate the acute takeoff angle. A critical component of this procedure relates to the definition of the size of the patch. Undersizing can result in insufficient relief of ostial stenosis, whereas oversizing may lead to kinking, thrombosis, or aneurysmal degeneration. Short-term results are excellent, with low operative risk, but long-term data are limited and likely depend on the durability of the patch material.[104]

CORONARY ARTERY BYPASS GRAFTING. CABG in patients with AAOCA is performed using the standard methods used for atherosclerotic coronary artery disease, including variations in access, use of cardiopulmonary bypass, and cardioplegic solutions. Among all the surgical solutions available to treat AAOCA in adults, CABG is technically the easiest.

The greater saphenous vein and the right and left internal thoracic arteries (in situ or anastomosed to the aorta) have been used as CABG conduits.[105-107] Arterial grafts probably have better long-term patency rates than venous grafts, but they are more vulnerable to the effect of chronic coronary competitive flow.[108,109]

A key and controversial issue is management of the AAOCA at the time of CABG. In most patients with AAOCA, the obstruction to coronary flow is intermittent, and there is a high risk of chronic competitive flow that is an important risk factor for CABG graft failure.[110] Ligation of the native coronary artery avoids coronary competitive flow and may potentially maximize graft patency, but it is not routinely adopted in clinical practice, probably out of concerns for rendering the coronary circulation totally dependent on graft flow in young and active patients. The evidence on CABG outcomes in patients with anomalous coronary arteries is limited to case reports and small cases series; the procedure is safe, with no reported operative mortality, but there are few long-term data (Supplemental Table 5). Cases of early graft failure and graft atresia when the native coronary artery was not ligated have been reported.[111,112] An important limitation of the available evidence is that all data on CABG graft patency and the effect of coronary competitive flow are derived from studies performed in patients with atherosclerotic coronary disease and may not be applicable to patients with AAOCA.

REIMPLANTATION. Coronary reimplantation requires mobilization of the proximal portion of the anomalous coronary that is transected.[113] The aortic wall is then closed (sometimes using a small pericardial patch), and the anomalous coronary is reimplanted in the anatomically correct aortic sinus. Care must be taken to avoid vessel trauma and kinking. The procedure can be technically challenging because the coronary artery wall can be very thin and frail, thus making direct anastomosis on the aorta difficult; in those cases, the use of a pericardial patch may be indicated. Coronary reimplantation may be combined with other techniques such as unroofing or ostioplasty.

PULMONARY TRANSLOCATION. Pulmonary translocation is aimed at enlarging the space between the great vessels to relieve compression on an AAOCA running between the aorta and the pulmonary artery.[114] The pulmonary artery is divided at its bifurcation, and a neobifurcation is created by anastomosing the main and left pulmonary arteries by using a patch.[115] Alternatively, the pulmonary artery can be moved anteriorly to the aorta.[115] The procedure in isolation is indicated only in the very uncommon cases where the interarterial course is the only anatomical abnormality thought to be associated with myocardial ischemia, but it may be associated with other interventions such as unroofing.

SURGICAL INTERVENTIONS FOR TRANSSEPTAL AAOCA. Two surgical options are available for transseptal AAOCA. In the posterior approach, the pulmonary artery is transected to gain access to the transseptal coronary artery and divide the overlying muscle.[116] This technique is most useful in cases where the transseptal component is superficial, and it has often been supplemented with an anterior translocation of the right pulmonary artery. An

alternative and more common technique is the transconal or transinfundibular unroofing, which is particularly indicated when the artery has a deep transseptal course.[117] In this technique, the right ventricular outflow tract is incised below the pulmonary valve, the transseptal coronary artery is unroofed through the posterior wall of the right ventricle, and a patch is sutured in the posterolateral part of the outflow tract.[41,118]

SELECTION OF THE SURGICAL TECHNIQUE. Given that ischemia in AAOCA may be caused by different mechanisms, surgical repair should address all of them, and a combination of different techniques may be necessary.

The decision on which surgical approach to use in patients with interarterial AAOCA is based on the presence, length, and location of the intramural segment. In case of a long intramural segment that travels above the level of the aortic valve, unroofing is the treatment of choice.[99] If the intramural segment travels below the level of the aortic valve, coronary reimplantation, ostioplasty, and partial unroofing are preferable options. If the intramural segment is short or if unroofing would place the ostium too close to the intercoronary pillar, coronary reimplantation should be considered.[36] In cases with no intramural segment and isolated stenosis of the coronary ostium that is located well away from the intercoronary pillar, ostioplasty is appropriate. Transseptal AAOCA is best treated by posterior unroofing if the segment is superficial or by transinfundibular unroofing if the intramyocardial segment is deep in the septum.

CABG should be reserved for patients with concomitant atherosclerotic coronary artery disease or when anatomical correction is suboptimal for technical or anatomical reasons.

Figure 6 presents a general guide for selection of the surgical technique.

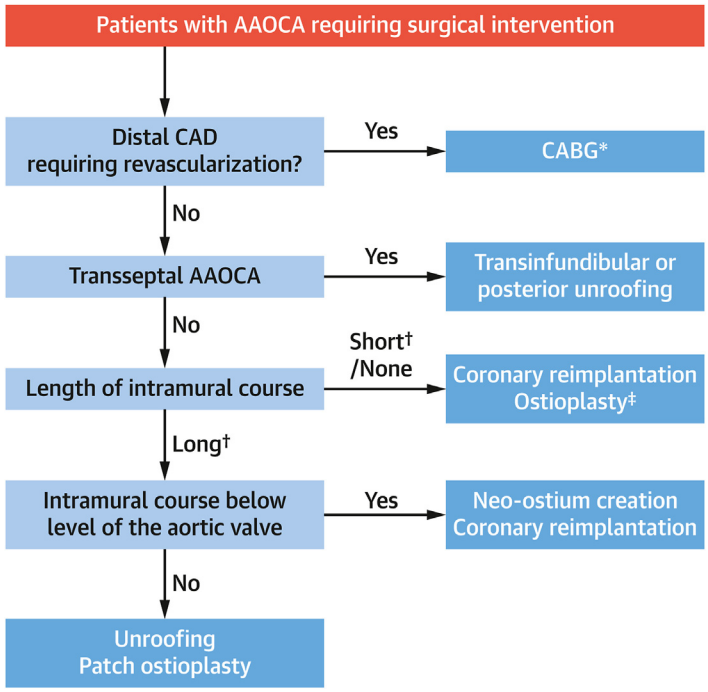


FIGURE 6 General guide for the selection of the surgical technique on the basis of key anatomical features. *CABG is also indicated when anatomic correction is considered suboptimal for technical or anatomic reasons. †A long intramural course refers to a course long enough to allow simple unroofing to virtually move the anomalous coronary to the correct sinus away from potential compression by the intercoronary commissure or pillar. ‡Simple ostioplasty may be considered in selected cases of isolated ostial stenosis with an anomalous coronary away from potential compression by a commissure or pillar. (AAOCA, anomalous aortic origin of the coronary arteries; CABG, coronary artery bypass grafting; CAD, coronary artery disease.)

PERCUTANEOUS TREATMENT OPTIONS

The evidence in support of percutaneous coronary interventions (PCIs) in AAOCA is limited to small case series performed both electively and in the acute setting.[119,120]

Percutaneous options may be aimed at treating either atherosclerotic coronary disease coexistent with AAOCA or the anomalous coronary artery segment itself (generally when a slit-like ostium or an intramural proximal segment is the key anatomical feature).[120]

Selecting an appropriate diagnostic or guiding catheter is the most important determinant of procedural success. An Amplatz left-type or multipurpose catheters are generally appropriate, but extra-backup can also be used. The right-sided anomalous circumflex artery may be challenging for stent delivery and can sometimes be treated with balloon angioplasty alone.[121] Guide extension catheters can offer support for successful delivery of stent catheters in challenging circumstances.

Drug-eluting stents with thick struts implanted with high pressures (≥20 bars) are used to optimize vessel remodeling when PCI is performed in the intramural segment of AAOCA. IVUS guidance is recommended.[119] In a study of 67 patients with AAOCA treated by PCI, the 5-year rate of in-stent restenosis was 13%.[76]

At present, the risk of elastic recoil after stenting of a thick arterial wall free of atherosclerosis and exposed to dynamic compression is largely unknown. Therefore, PCI should be restricted to nonsurgical candidates until more evidence becomes available.

EXPERT STATEMENTS AND SUPPORTIVE TEXT

- In patients with AAOCA, shared decision making considering all the available evidence,

individual patient characteristics, and preference is recommended.

Given the current key gaps in knowledge on AAOCA, shared decision making including the patient and all involved health providers is critical to individualize treatment to the patient's needs and expectations.

- Correction of AAOCA in adult patients should be performed at centers with experience in all the available approaches.

Correction of AAOCA in adults can be performed using approaches encompassing a wide range of technical difficulty, some of which may be familiar only to centers with specific expertise. It is important that the choice of technique for intervention is based on the coronary anatomy and the patient's clinical status rather than on the locally available options.

- Surgical repair should address all potential mechanisms of ischemia, and a combination of different techniques may be necessary.

Ischemia in patients with AAOCA may result from different mechanisms of fixed and dynamic stenosis or compression of the coronary artery. Surgical repair should address all potential mechanisms.

- Anatomical repair should be preferred when possible. Coronary bypass surgery may be considered when anatomical repair is not possible or has a low probability of success or when there is concomitant coronary artery disease.

Coronary bypass surgery is a simple intervention with low operative risk. However, its efficacy in patients with AAOCA has been poorly studied, and there are concerns about long-term graft patency.

- When coronary bypass surgery is performed, the native coronary artery should be ligated if possible, and arterial, rather than venous, grafts should be used.

When coronary bypass surgery is performed in patients with AAOCA, it is key that the chances of long-term graft patency are maximized. Elimination of coronary competitive flow and use of arterial grafts are important technical modifications to achieve this goal. If possible, ligation of the native coronary artery should be performed after a test period of temporary occlusion with verification of good graft flow and no evidence of ischemia. The use of aorta-anastomosed arterial grafts may provide higher immediate flow and reduce concerns for competitive flow and ischemia when ligating the native artery, but there is limited available evidence.

- In asymptomatic patients with no evidence of myocardial ischemia, the presence of high-risk anatomy and the chances of a successful anatomical repair should influence the decision to intervene.

For asymptomatic patients with no evidence of myocardial ischemia (in particular when the RCA is involved) the chances of anatomical repair should be weighted in the clinical decision making.

- In most AAOCA patients undergoing noncoronary surgery, concomitant repair of the coronary anomaly should be considered.

In most cases, adding repair of AAOCA to the primary cardiac operation does not increase the operative risk, and it may have long-term benefits. The decision to intervene must be based on the type of coronary anomaly, the risk-to-benefit ratio, and the chance of anatomical repair in each individual patient.

- Percutaneous treatment of AAOCA should be restricted to nonsurgical candidates.

When AAOCA treatment is indicated, there currently is very limited evidence to support percutaneous options.

- Future research is urgently needed to determine the prevalence, prognosis, and natural history of AAOCA, define the utility of imaging techniques, and compare the results of different treatments.

Current evidence is largely inadequate to inform clinical decision making, and more research is needed.

Details of agreement for each statement are reported in the Supplemental Appendix.

GAPS IN KNOWLEDGE AND FUTURE DIRECTIONS

Critical gaps remain in our knowledge of AAOCA. Future research is urgently warranted to better define the prevalence, prognosis, and natural history of AAOCA, as well as to assess the utility of different imaging techniques, determine best strategies for risk stratification, and establish the efficacy of different treatments. Given the relative rarity of AAOCA and the anatomical and clinical variability, international collaboration will be critical to shed light on current uncertainties.

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