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INVITED EXPERT REVIEWS

Anomalous Aortic Origin of a Coronary Artery

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ABSTRACT

BACKGROUND Although anomalous aortic origin of a coronary artery (AAOCA) is associated with risk of sudden cardiac arrest (SCA), there is a spectrum of disease, with the appropriate management for many remaining unclear. Increasing data warrant review for an updated perspective on management.

METHODS A panel of congenital cardiac surgeons, cardiologists, and imaging practitioners reviewed the current literature related to AAOCA and its management. Survey of relevant publications from 2010 to the present in PubMed was performed.

RESULTS The prevalence of AAOCA is 0.4% to 0.8%. Anomalous left coronary artery is 3 to 8 times less common than anomalous right coronary, but carries a much higher risk of SCA. Nevertheless, anomalous right coronary is not completely benign; 10% demonstrate ischemia, and it remains an important cause of SCA. Decision-making regarding which patients should be recommended for surgical intervention includes determining anatomic features associated with ischemia, evidence of ischemia on provocative testing, and concerning cardiovascular symptoms. Ischemia testing continues to prove challenging with low sensitivity and specificity, but the utility of new modalities is an active area of research. Surgical interventions focus on creating an unobstructed path for blood flow and choosing the appropriate surgical technique given the anatomy to accomplish this. Nontrivial morbidity has been reported with surgery, including new-onset ischemia.

CONCLUSIONS A proportion of patients with AAOCA demonstrate features and ischemia that warrant surgical intervention. Continued work remains to improve the ability to detect inducible ischemia, to risk stratify these patients, and to provide guidance in terms of which patients warrant surgical intervention.

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Anomalous aortic origin of a coronary artery (AAOCA) describes a coronary that arises from the aorta at a site other than the appropriate coronary sinus. The management of AAOCA remains controversial. The aim of this expert review is to collate and critically review the most recent data for an updated perspective by experts in the field. For the purposes of this surgical journal, the focus is on decision-making and surgical techniques, with further details of other aspects contained in the Supplemental Material.

PATIENTS AND METHODS

Publications written in English in PubMed were reviewed by relevant key words (AAOCA, anomalous coronary artery); studies from 2010 to the present were

The Supplemental Material can be viewed in the online version of this article [https://doi.org/10.1016/j.athoracsur.2024.01.016] on http://www.annalsthoracicsurgery.org.

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surveyed. Case reports were excluded. Emphasis was given to larger cohorts and studies with clinical and imaging follow-up.

BACKGROUND

The exact prevalence of AAOCA is challenging to determine, with the population presenting for imaging or at autopsy usually not representative of the general population. With these limitations recognized, the estimated prevalence is approximately 0.4% to 0.8%,[1,2] with anomalous aortic origin of the right coronary artery (R-AAOCA) 3 to 8 times more common than that of the left (L-AAOCA).[3,4] Natural history analyses center around the risk of sudden cardiac arrest (SCA). This is challenging to quantify, given the infrequency of the event of interest (SCA) requiring a large number of patient-years to detect, and the uncertainty about the prevalence of AAOCA (ie, the denominator). Furthermore, evidence continues to mount that risk varies dependent on anatomic features, and such details are usually not available. Risk of SCA is associated with exertion, notably in those with interarterial L-AAOCA,[5] and appears to be age related, with most occurring in patients aged 10 to 30 years.[6,7] However, there does not seem to be an age cutoff beyond which AAOCA may no longer be relevant.[8-10] Analysis using the data of Maron and Brothers calculated the cumulative risk of death for those aged 15 to 35 years in sports to be 6.3% for L-AAOCA and 0.2% for R-AAOCA.[11-13] Further details of AAOCA prevalence, genetics and embryology of AAOCA, pathophysiologic mechanism behind SCA, and risk of SCA are included in the Supplemental Material.

IMAGING OF AAOCA

Imaging is fundamental not only to the diagnosis of AAOCA but also to risk stratification and decision-making. Computed tomography angiography (CTA) has been the primary modality for this purpose, but other modalities, such as cardiac magnetic resonance, can be useful. A synopsis is included here, and further details on the diagnosis of AAOCA and imaging are included in the Supplemental Material.

Key to imaging is evaluation for anatomic “high-risk” features of AAOCA, which are important for management decision-making. These include slitlike orifice, stenotic orifice, acute angulation, and narrowed elliptical proximal coronary shape,[4,9,14-16] all of which relate to the coronary artery’s intramural course (course of the proximal coronary within the aortic wall). Properties of the distal coronary, including the presence of myocardial bridges and atherosclerotic disease, should also be evaluated. Myocardial bridges can cause significant symptoms and have been reported as a reason for reoperation,[17-19] as has SCA after unroofing when the bridge was not recognized and addressed.[20] In adults with AAOCA, accelerated atherosclerosis has been seen in anomalous left circumflex with a posterior course,[21] but no clear increased atherosclerotic coronary artery disease burden has been reported in other types of AAOCA.[22]

Given the variation in morphologic appearance and the impact that features have on both risk stratification and choice of surgical technique, a unified language and classification system was needed. This would aid multidisciplinary discussions and assessment of appropriate surgical technique and allow comparisons across centers. To this end, Mery and colleagues[14] have developed a topography nomenclature system, and a structured reporting template for AAOCA has been published (see Supplemental Material).[23]

EVALUATION FOR ISCHEMIA

A key tenant for evaluating patients with AAOCA and stratification of their risk, thereby informing the decision to recommend surgical intervention, is whether there is evidence of ischemia. Such evaluation is challenging and continuously evolving. The American Association for Thoracic Surgery guidelines recommend that an exercise test be combined with nuclear perfusion scan or stress echocardiography.[1] Stress testing is useful when the result is positive, but it offers limited reassurance when the result is negative, given the low negative predictive power.[1] Considerable work in this area has been performed since the publication of those guidelines as well as of the American Heart Association/American College of Cardiology and European Society of Cardiology adult congenital heart disease guidelines[9,24] that can help inform evaluation for ischemia as covered in this section (further details in the Supplemental Material). In the search for more sensitive provocative tests to assess for ischemia, certain institutions have turned to dobutamine stress cardiac magnetic resonance as a promising modality, and it can be used in pediatric patients safely and effectively.[25] In some practices, often those connected to adult patients, dobutamine positron emission tomography is used to evaluate for ischemia.

Catheter-based physiologic assessment is being investigated for use in the AAOCA population. One challenge is the different anatomy and pathophysiology in the AAOCA population compared with patients with atherosclerotic disease and what should be considered the cutoff for high risk or indicative of ischemia. Given the dynamic nature of obstruction in AAOCA and the specific parameters used in instantaneous wave-free ratio (iFR) measurements, iFR has become a potentially useful tool for evaluating these patients.[26] Several institutions have used iFR with dobutamine provocation to mimic physiologic stress, with ≤0.85 at rest or with inotropic stress supporting a significant blood flow compromise to the myocardium. Intravascular ultrasound can be a complementary tool to demonstrate anatomic information not always gleaned by noninvasive imaging or coronary angiography, including slitlike orifice anatomy, luminal area of the ostium, extent of dynamic compression, and any plaque—all features that can affect ischemia.

There is a need to expand on such approaches to distinguish AAOCA patients with and without clinical ischemia. At present, the best approach to evaluating ischemia in AAOCA is a systematic and multimodality approach that incorporates presenting symptoms, comorbidities such as obstructive coronary artery disease, noninvasive provocative testing, and potentially selective invasive ischemia evaluations.

RECOMMENDATIONS FOR SURGICAL INTERVENTION

The recommendation for surgical intervention is nuanced, with gray areas remaining to be defined. The 2017 AAOCA expert consensus guidelines[1] recommend exercise restriction and referral for surgery for those with SCA, for those with symptoms of ischemic chest pain or syncope suspected to be due to ventricular arrhythmias, and for all patients with L-AAOCA from the right sinus and interarterial course (class I). Importantly, those with L-AAOCA should still undergo evaluation for ischemia unless they presented with SCA. The indications for surgery for R-AAOCA, listed as class IIa, include those with symptoms concerning for ischemia or a positive exercise stress test result. The 2018 adult congenital heart disease guidelines are largely similar, with pertinent differences being that L-AAOCA without ischemic symptoms or evidence of ischemia is a class IIa recommendation for surgery. For R-AAOCA, the recommendations are also similar, but ischemic symptoms or ischemia on workup is a class I recommendation for surgery, no ischemic symptoms or evidence of ischemia on workup but ventricular arrhythmia is a class IIa, and then none of the preceding is a class IIb. The 2020 European Society of Cardiology guidelines are more specific and recommend surgical intervention in symptomatic patients with either evidence of stress-induced myocardial ischemia in the affected territory or high-risk anatomy (class I). They recommend consideration of surgical intervention (class IIa) in asymptomatic patients if there is evidence of ischemia or if they have L-AAOCA with high-risk anatomy. Surgery may be considered (class IIb) in symptomatic patients without ischemia or high-risk anatomy or in asymptomatic patients with L-AAOCA and no ischemia or high-risk anatomy when they present <35 years old.[24]

Whereas these recommendations are helpful, many cardiologists and surgeons are faced with patients with symptoms that are not clearly ischemic and an exercise test result that is negative for ischemia. Furthermore, the patient and family's interaction with the diagnosis and its implications (including fear of SCA, athletic aspirations) and concerns related to surgical intervention play important roles in and have an impact on shared decision-making.

As in any decision to perform an intervention, the risks and benefits of the intervention must be weighed against the risks and benefits of not intervening. Differentiating whether symptoms are secondary to ischemia is key in decision-making. To this end, improved methods for detecting ischemia during exercise in this population, as discussed before, will be extremely valuable. In the meantime, we take into consideration high-risk anatomic features. As described in the European Society of Cardiology guidelines,[24] these include an intramural course and orifice anomalies (slitlike orifice, acute angle takeoff, orifice >1 cm above the sinotubular junction). Whereas intramural course for both L-AAOCA and R-AAOCA has been necessary for some institutions to recommend surgical intervention, in the Congenital Heart Surgeons' Society (CHSS) cohort, 12% with ischemia did not have an intramural course.[16] Although these high-risk features have been identified as having more risk relative to other variants, we do not currently have data clarifying which feature or group of features would pose such risk without intervention, particularly in the R-AAOCA subtype, that surgical intervention offers a clear benefit. Analyzing whether there is a net benefit for surgical intervention compared with nonsurgical intervention in the early, mid, or long term is also dependent on the specific surgery performed and surgeon/institutional procedure-specific experience.

The goal of surgery is reducing risk of SCA and ischemic symptoms. Data suggest that SCA risk is greatest in the 12- to 22-year age bracket, likely owing to competitive sport participation in this age range. Whereas SCA is the most important consequence of AAOCA to be addressed with surgery, the risk of SCA is not completely eliminated as SCA after surgery has been reported.[27,28] Another caveat in the risk-benefit assessment of surgery is whether exercise restriction is being recommended if surgery is not performed. However, exercise restriction is generally not recommended unless it is short term pending further evaluation or surgery.[29]

Symptom relief with surgery is variable. Meijer and colleagues[30] described 53 adolescents and adults with AAOCA with a median age of 44 years. 97% were symptomatic before surgery, with 35% having angina-type symptoms; after surgery, 59% were symptom free,

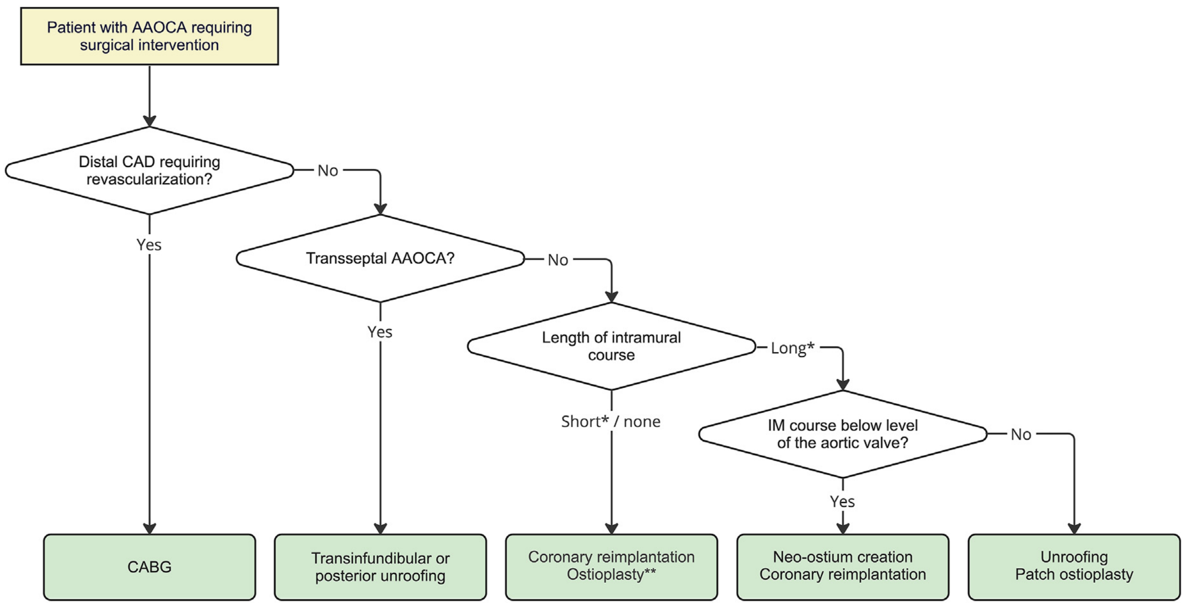

FIGURE 1 Decision-making in anomalous aortic origin of a coronary artery (AAOCA). *Long (vs short) intramural segment is one in which surgical unroofing allows placement of the coronary ostium in the correct sinus away from potential compression by the intercoronary pillar. **Ostioplasty may be considered in cases with isolated ostial abnormalities. (CABG, coronary artery bypass grafting; CAD, coronary artery disease; IM, intramural.) Reprinted with permission by Carlos Mery and Dell Medical School © 2023.

with 3 patients (9%) presenting with angina and requiring reoperation.[30] Wittlieb-Weber and colleagues[31] also examined symptoms after surgery in a cohort of 24 pediatric patients with a median age at follow-up of 18 years. They found that 54% still had cardiac symptoms, mostly chest pain that was not found to be ischemic on workup; as well, roughly half had symptoms similar to before surgery. In the recently published Mayo cohort of 148 patients,[8] 96 patients experienced ischemic symptoms before repair and 36 had symptoms after surgery, with no evidence of ischemia on workup. These data, however, are in contrast to the series by Mery and coworkers[14] of 44 patients, in which 91% were asymptomatic at last follow-up with a median follow-up of 2 years, and by Mainwaring and coworkers,[19] in which 97% of the patients who were symptomatic preoperatively were free of symptoms postoperatively.

We have recently accumulated more data on the early and midterm risks related to surgical intervention and which types of anatomy pose a lower risk surgical substrate; discussion is ongoing as to whether certain surgical techniques may be better in certain anatomic situations. Whereas certain single-center case series have reported favorable results of surgery, other studies including the multicenter CHSS cohort suggest that significant risk and morbidity can occur with surgical intervention. As always, these data must be interpreted in the context of a given institution and surgeon, in consideration of the clinical condition and anatomy of the patient, and in discussion of the concerns of the family and the patient.

AAOCA can also be an incidental finding in those with congenital heart disease. Anomalous coronaries in the setting of truncus and transposition are outside of the scope of this document. Whether an anomalous coronary artery should be addressed at the time of other types of congenital heart surgery should be considered on a case-by-case basis and generally should be addressed if the patient meets indications for surgery on the coronary independent of other lesions. The size and age at which performing congenital heart disease repair concomitant with an AAOCA repair is reasonable should be a case-by-case decision, with intervention likely to be reserved for older children and teenagers unless there is evidence of ischemia.

SURGICAL TECHNIQUES

The goal of repair is to ensure that blood traveling to the myocardium does not encounter obstruction due to ostial stenosis, compression by the commissural pillar, acute angulation exiting the aorta, pathway between the great vessels, intramural course, and compression due to an intramyocardial course (transseptal or myocardial bridge).[32] A number of surgical techniques have been described, with techniques varying both by institution and by anatomy. Given the array of employable

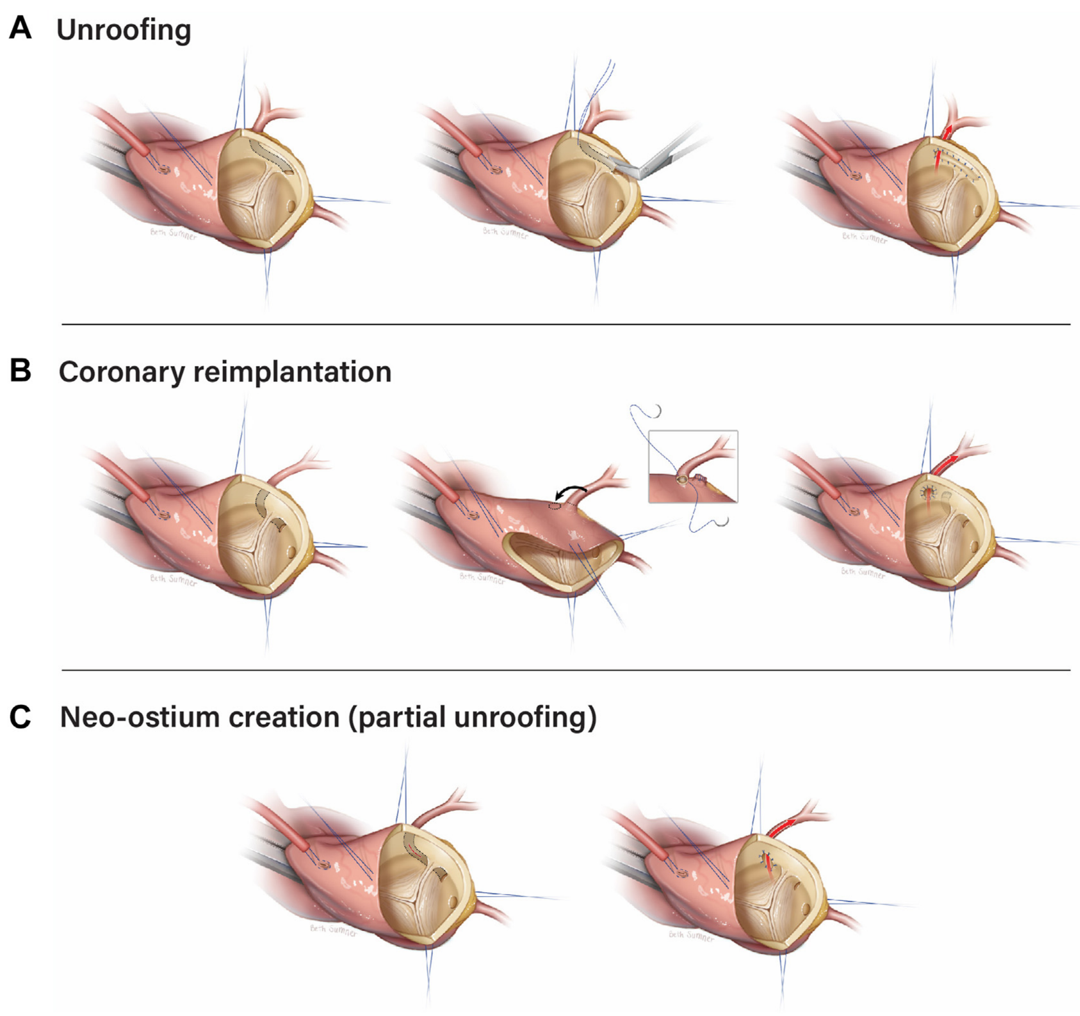

FIGURE 2 Illustration of surgical techniques in the case of anomalous aortic origin of the left coronary artery with intramural course ghosted in gray. (A) Unroofing technique; note that the intramural course remains above the commissural post. Potts scissors are shown dividing the overlying fibrous tissue (middle image); in the completed repair (right image), the red arrow demonstrates unobstructed blood flow from the aortic lumen into the distal portion of the left coronary. Tacking sutures are shown securing the edges of the unroofed segment. (B) Reimplantation technique; note that the intramural segment runs inferior to the top of the commissural post. The coronary artery is transected where it leaves the aortic wall and transferred (arrow) to the new location in the center of the left sinus (inset). This is completed with a running, fine Prolene suture. The finished result (right image) shows unobstructed blood flow from the aorta into the distal coronary artery. (C) “Neo-ostium” technique; note that the intramural segment runs inferior to the top of the commissural post. The aortic wall is incised (red line, left image) overlying the portion of the left coronary artery as it exits the aortic wall, that is, the distal intramural segment. The edges of this new ostium are tacked down (middle image), and unobstructed blood flow (red arrow, right image) from the left sinus transits into the distal coronary artery. Illustration © 2023 Ascension. Used with permission.

techniques, the anatomy should be matched with the appropriate technique as well as with the surgeon’s experience (Figure 1).

The most widely used technique, “unroofing” (Figure 2A), involves dividing the portion of the aortic wall overlying the intramural segment and tacking the fibrous tissue. When this path disturbs the aortic valve commissural post, the post is commonly tacked to the aortic wall to prevent aortic insufficiency. Critical to this technique is ensuring that the coronary is completely unroofed and the new opening for the coronary does not have a residual angulation or course behind the aortic valve intercoronary pillar where it can be compressed.[33] In cases in which unroofing will position the new orifice in the appropriate sinus and the orifice will no longer be

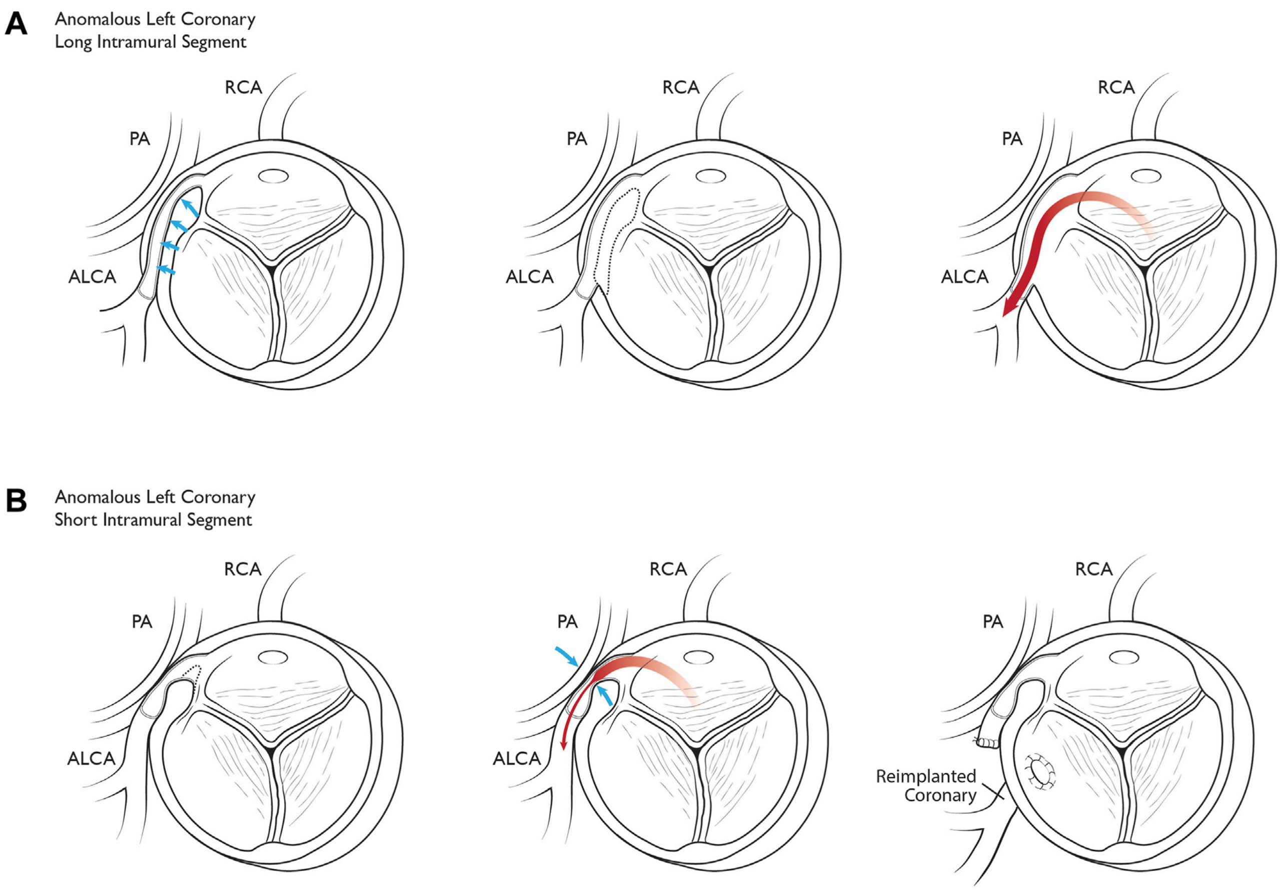

FIGURE 3 The unroofing procedure in the setting of (A) long and (B) short intramural segments. (A) If there is a long intramural segment, unroofing eliminates the intramural segment and virtually places the ostium of the unroofed coronary in the correct sinus, thus correcting all potential culprits of compression (sinus of origin, intramural segment, interarterial segment, and ostial abnormalities). (B) If the intramural segment is short, unroofing may eliminate the intramural segment and improve the ostial morphology, but the coronary may arise close to the intercoronary pillar and be at risk for compression. An alternative strategy, such as reimplantation, may be preferable. (ALCA, anomalous left coronary artery; PA, pulmonary artery; RCA, right coronary artery.) Adapted from Mery and colleagues[14] with permission by Texas Children's Hospital © 2016.

interarterial, this can be an appropriate surgical strategy. In cases in which a short course exists, unroofing may leave the new orifice juxtacommissural and interarterial, both of which may compromise coronary blood flow. In these cases, different surgical techniques are recommended (Figure 3).

One concern related to unroofing when the coronary runs posterior to the commissural pillar is that takedown and resuspension of the post can be associated with aortic insufficiency.[34] This has led to enthusiasm regarding "neo-ostium" creation (discussed later), aortocoronary window creation, and reimplantation with or without a button. Conceptually, these techniques have the advantages of creating a wide-open ostium in the appropriate sinus without disrupting the aortic valve. The reimplantation technique (Figure 2B) does require significant dissection external to the aorta, and care must be taken to not kink the coronary artery. Judging length can be challenging while on bypass. Whereas surgeons regularly performing root surgery may be more comfortable with this technique compared with those not regularly performing root surgery, this is more technically challenging than root surgery in that the reimplantation is generally performed using the coronary wall as opposed to the button of aortic wall tissue. Surgeons employing the reimplantation technique recognize the risks related to the procedure,[35] but several think this may give a more anatomic result that if done correctly may have a better long-term outcome. This technique can also be useful if there is a short intramural course, in which unroofing would leave the ostium in the

TABLE Outcomes of Surgery for AAOCA

First Author	Year	No. of Patients	Single Center vs Multicenter	Age (y)	Preoperative Symptoms/ Ischemia	RCA, LCA	Techniques	Complications	Follow-up	Postoperative Ischemia	Postoperative Symptoms	Comments
Sharma[43,a]	2014	75	Single center	40	72% angina, shortness of breath, or syncope 53% had stress testing, and results were abnormal in 50%	92% RCA 8% LCA	Unroofing in all	2 had CABG for flow acceleration at RCA origin on TEE early in experience 2 atrial fibrillation 1 pericarditis 1 subdural hematoma in 57-year-old redo AVR unroofing of AAORCA who later died in rehabilitation center	18 months	None	No recurrent symptoms	13% associated procedures
Fabozzo[44]	2016	72 surgical	Single center	12	75% symptomatic; 5 had evidence of ischemia on workup	72% RCA 28% LCA 5 intraconal	Unroofing 86% Reimplantation 11% 2 myocardial bridge	4 patients; 1 early AR, 1 ECMO for coronary spasm, reoperation from outside hospital for AAOCA, 1 mediastinitis, and 1 late AV repair	1.4 years	53% postoperative evaluation for ischemia, all negative	3 chest pain with pericarditis 12 nonexertional chest pain and ischemia workup negative	42% of RCA had repair, 71% of LCA Surgery was associated with LCA, age >10 years, exercise restriction on univariate analysis; on multivariate analysis, association with chest pain or syncope and older age
Mainwaring[19]	2016	115	Single center	16	51% symptoms consistent with ischemia	66% RCA 32% LCA	Unroofing 75% Reimplantation 8% PA translocation 17%	1 early revision 4 postcardiotomy syndrome 1 transient heart block 1 with myocardial damage preoperatively ultimately underwent transplantation	6 years	2 with myocardial ischemia, both reoperation for bridges	97% of the symptomatic patients were free of symptoms 2 had recurrent symptoms and had reoperation	
Balasubramanya[39]	2017	66	Single center	12	79% symptomatic 30% anginal	83% RCA 17% LCA	Unroofing 89% Neo-ostium 8% Side-to-side 3%	Thrombus in coronary in 1 patient Pericardial effusion 4 months later in 1 patient	5.6 years	None	Not detailed	Symptoms did not correlate with length or diameter
Nees[45]	2018	60	Single center	15	63% chest pain	50% RCA 50% LCA	93% unroofing 7% translocation	10% postoperative complications including transient atrial arrhythmia, superficial wound infection, pneumothorax Postdischarge 18% postpericardiotomy syndrome 6% had moderate AR on follow-up and 2% severe AR	1.6 years 90% complete	57% had exercise stress 6% evidence of ischemia, additional 2 (11%) abnormal nuclear	60% complete resolution 17% change No new symptoms for those asymptomatic	

(*Continued*)

TABLE Continued

First Author	Year	No. of Patients	Single Center vs Multicenter	Age (y)	Preoperative Symptoms/ Ischemia	RCA, LCA	Techniques	Complications	Follow-up	Postoperative Ischemia	Postoperative Symptoms	Comments
Sachdeva[28]	2018	63	Single center	13	50% symptoms possible ischemia	79% RCA 21% LCA	All unroofing	No perioperative complications 1 required AVR 6 months later	3.1 years	No ischemia identified on testing, but 3 had SCA; 2 of these had aborted SCA before surgery	Symptoms persisted or developed in 46%	
Padalino[18]	2019	156 surgical	Multicenter	39	87% symptomatic with 66% exertional 23 of 69 exercise stress test positive, 10 of 16 nuclear	67% RCA 22% LCA	Unroofing 56% Reimplantation 19% CABG 15%	1% mortality 5% early reoperation 4% MCS 4% effusions 3% arrhythmia	18 months	3 reoperations 3 stents 1 myocardial bridge repair Testing for ischemia performed in minority, 1 positive stress test result	Not detailed	
Gaillard[35]	2020	61	Single center	15	70% symptomatic 10 positive exercise stress test, 6 positive nuclear, 2 positive cMR	66% RCA 33% LCA 5 intraseptal	57% "anatomic repair" 31% coronary translocation 5 intraseptal had release of artery from septum	5% acute postoperative ischemia	38 months	2 patients needed late coronary intervention 3 patients had patch aneurysm 50% had testing for ischemia, all except 1 negative	All asymptomatic except 1	
Jegatheeswaran[34]	2020	395 surgical	Multicenter	13	163 had preoperative ischemia testing 64 had evidence of ischemia	71% RCA 27% LCA	Unroofing 87% Reimplantation 6%	Composite adverse perioperative events 7%-13%, depending on anatomy and technique	2.8 years	80% no longer had evidence of ischemia 13 new ischemia	Not detailed	Increased risk subgroups included preoperative ischemia, AAOLCA, repair other than unroofing, and commissural manipulation
Bonilla-Ramirez[41]	2021	61	Single center	15	54% exertional symptoms 8% abnormal sNPI 25% abnormal sCMR	84% RCA 16% LCA	Unroofing 72% Reimplantation 28%	13% perioperative complications 1 coronary stenosis requiring CABG 8% pericardial effusion	4 years	1 recurrent SCA due to myocardial bridge 6 of 42 found to have inducible ischemia	9% nonexertional symptoms 3% exertional symptoms/ ischemia	
Doan[46]	2023	52 surgical	Single center	11	39% exertional symptoms 33% inducible hypoperfusion	All RCA	Unroofing 73% Reimplantation 27%	1 CABG because of technical difficulty No details on pericarditis and other perioperative complications	5.5 years	4 abnormal stress perfusion at 3 months, subsequent dobutamine MR without hypoperfusion	1 nonexertional chest pain	
Patlolla[8]	2023	148	Single center	44	65% ischemic symptoms; of these, 48% had evidence of ischemia	88% RCA 12% LCA	All unroofing	1 readmit mediastinitis	9.5 years	None	24% chest pain, none ischemic	20% concomitant procedures

(Continued)

TABLE Continued

First Author	Year	No. of Patients	Single Center vs Multicenter	Age (y)	Preoperative Symptoms/ Ischemia	RCA, LCA	Techniques	Complications	Follow-up	Postoperative Ischemia	Postoperative Symptoms	Comments
Pregaldini[47]	2023	71	2 centers	55	93% "symptomatic" No documentation of ischemia	79% RCA 15% LCA	Unroofing 72% Reimplantation 28% Neo-ostium patchplasty 13%	4% MI and underwent CABG 9% MCS	3.1 years 89% complete	3 patients requiring early reintervention	70% asymptomatic, 18% no symptom improvement; of these, no evidence of ischemia	39% concomitant procedures

[a]These patients are also included in the study by Patlolla[8] from 2023. This table includes series with >50 patients undergoing surgery. AAOCA, anomalous aortic origin of coronary artery; AAOLCA, anomalous aortic origin of left coronary artery; AAORCA, anomalous aortic origin of right coronary artery; AR, aortic regurgitation; AV, aortic valve; AVR, aortic valve replacement; CABG, coronary artery bypass graft; cMR, cardiac magnetic resonance; ECMO, extracorporeal membrane oxygenation; LCA, left coronary artery; MCS, mechanical circulatory support; MI, myocardial infarction; MR, magnetic resonance; PA, pulmonary artery; RCA, right coronary artery; SCA, sudden cardiac arrest; sCMR, stress cardiac magnetic resonance; sNPI, stress nuclear perfusion imaging; TEE, transesophageal echocardiography.

inappropriate sinus or near the commissural post at risk for obstruction.

The neo-ostium creation technique (Figure 2C) can also be employed in cases in which there is a long intramural segment and the aortic valve would be disturbed by unroofing. In this case, a "keyhole" or neo-ostium is created within the correct anatomic sinus where the intramural portion of the coronary ends and the coronary exits the aortic wall. This, then, is a "partial" unroofing, such that the new ostium creates a direct entry of blood from the aorta into the coronary artery without any angulation.

Osteoplasty involves patch augmentation to enlarge the ostium and can be used with unroofing. This is frequently employed when there is residual narrowing as the coronary exits the intramural segment and leaves the aortic wall. For this technique to be performed, the aorta above the coronary is incised and a patch, frequently a triangular piece of autologous pericardium, is sutured to augment this area and to eliminate any acute angle or narrowing.

Pulmonary artery translocation has been used by certain groups as an adjunctive technique, given the role that compression between the pulmonary artery and aorta may play in ischemia.[19]

Surgical management of a transseptal coronary can include a posterior approach in which the pulmonary artery is transected and overlying muscle removed. This is most appropriate for a coronary that is relatively superficial, and some have augmented the technique with anterior translocation of the pulmonary root.[36] The second is an infundibular approach, in which the coronary is approached below the pulmonary valve and completely unroofed, and then a patch is placed on the right ventricular outflow tract.[37]

Coronary artery bypass is not considered the primary procedure of choice to treat AAOCA, except in the setting of atherosclerotic disease in the anomalous coronary. Because the coronary obstruction in AAOCA is dynamic in nature, the chronic competitive flow puts bypass grafts, particularly mammary arteries, at risk for graft failure. Graft failure has been reported both perioperatively and on follow-up,[38] but data are limited. Coronary artery bypass grafts may be useful when there is concern for coronary obstruction after the AAOCA repair.

EARLY OUTCOMES OF SURGERY

A number of single-center case series have been published from centers with a high volume of AAOCA (Table). Short-term outcomes are overall good, but there are potential complications. In a series of 66 patients, Balasubramanya and coworkers[39] reported 2 notable complications: 1 pericardial effusion requiring drainage

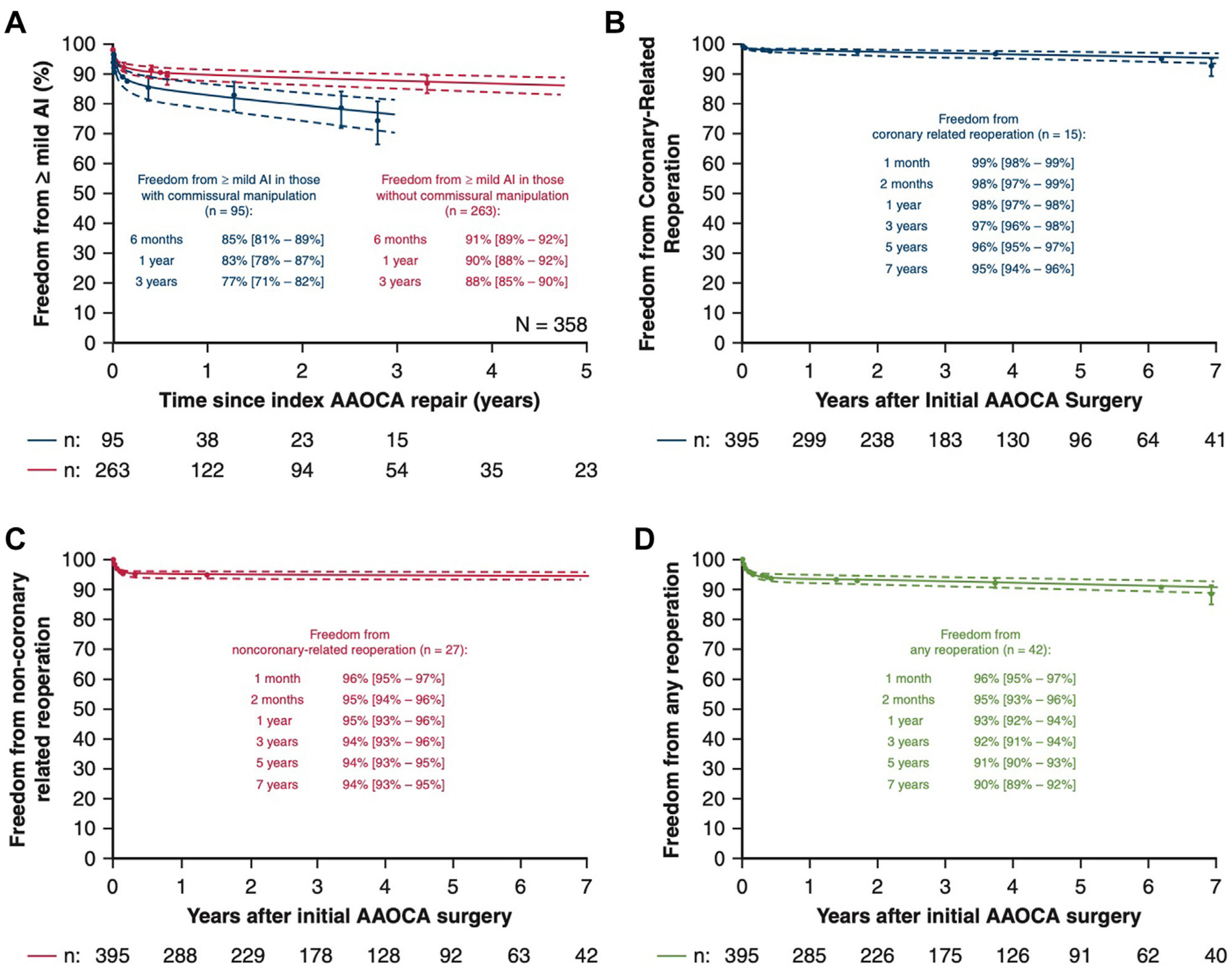

FIGURE 4 Kaplan-Meier curves. (A) Freedom from mild or more severe aortic insufficiency (AI) stratified by commissural post manipulation. (B) Freedom from coronary-related reoperation. (C) Freedom from non–coronary-related reoperation. (D) Freedom from any reoperation. Number of patients remaining at risk is shown on the x-axis. (AAOCA, anomalous aortic origin of coronary artery.) Reprinted from Jegatheeswaran and colleagues[34] with permission from The American Association for Thoracic Surgery.

4 months after surgery and 1 clot to the right coronary artery leading to surgical exploration. In a series of 44 patients, Mery and coworkers[14] reported that 9% of patients had pericardial effusions and 80% experienced pericarditis. In addition, 5 patients (12%) had mild persistent narrowing of the coronary artery on CTA after surgery; 4 of them were patients who had undergone unroofing of short intramural segments. In a study by Mainwaring and coworkers[19] including 115 patients, 2 had recurrent symptoms and underwent reoperation including 1 revision of the repair and 1 repair of myocardial bridge, 4 had pleural effusions, 4 had postcardiotomy syndrome, and 1 had transient heart block. In the recent Mayo cohort of 148 patients[8] with a median age of 44 years, there were no major complications, and 1 patient is being observed for mild neo-ostial narrowing. There was 1 complication after discharge in a patient readmitted with mediastinitis. Jiang and colleagues,[40] examining AAOCA in adults, found that 22% (26 patients) had AAOCA addressed at time of surgery with few perioperative complications, including new-onset atrial arrhythmias in 12%. Gaillard and colleagues[35] described 61 patients including 37 who underwent patch osteoplasty and 19 who underwent reimplantation. Three patients had postoperative coronary complications. During 38-month follow-up, 1 patient had ostial stenosis and 3 patients had patch aneurysm. Of 30 patients with postoperative testing for ischemia, 1 was positive for ischemia.

Bonilla-Ramirez and coworkers[41] described 16 patients who underwent reimplantation and compared those patients with 45 patients who underwent unroofing. Reimplantation was used when the coronary coursed below the commissure or if unroofing would result in the ostium residing in the inappropriate sinus or in a location at risk for compression by the intercoronary pillar. One of the 16 patients required coronary artery bypass grafting for postoperative ischemia.

The CHSS cohort, which enrolls patients ≤30 years old with AAOCA, has been invaluable in providing a more real-world look at the patients undergoing surgical repair of AAOCA. In the 2020 report by Jegatheeswaran and colleagues,[34] 395 patients underwent surgery at 45 centers, with 87% undergoing unroofing. Median follow-up was 2.8 years. Probability of operative death was 1% (n = 4), and 13 patients (3%) underwent reoperation for coronary issues, yielding a 5-year coronary-related reoperation rate of 4%; 80% no longer had ischemia, but 13 patients (4%) had new ischemia. Fourteen patients (4%) had postcardiotomy syndrome requiring medical therapy. In the nonsurgical group, there was a 1% sudden death rate during follow-up. When the combined adverse events of new moderate or more severe aortic regurgitation, new abnormal ejection fraction, any positive ischemia test result or symptoms after surgery or need for a coronary-related reoperation, requirement for extracorporeal membrane oxygenation, and death were considered, the incidence was 9% (34 patients). This analysis also found that surgical strategy other than unroofing had a significantly higher risk of complications (12%-17% vs 5%-9%).[34] Mild aortic regurgitation tended to be higher in those with commissural post manipulation (23% vs 12% at 3 years; $P = .05$). Figure 4 demonstrates the Kaplan-Meier curves for freedom from mild or more severe aortic regurgitation, any reoperation, and coronary artery-related reoperation.

An additional study of the CHSS cohort examined patients with ischemia[16]; 49 patients had ischemia, with a prevalence of 10% in R-AAOCA and 38% in L-AAOCA. Features associated with ischemia in L-AAOCA included an intramural course, high origin, and slitlike takeoff; features of R-AAOCA related to ischemia included a longer intramural course. Of note, 12% of patients with ischemia did not have an intramural course. The most common mode of presentation for ischemia patients (both L-AAOCA and R-AAOCA) was SCA. This study highlights that R-AAOCA may not be as benign as thought, with half of deaths (6/13) in the R-AAOCA group and 10% having evidence of ischemia on testing.

Several meta-analyses have been reported in AAOCA. Karangelis and coworkers[38] analyzed 7 studies of adults with AAOCA undergoing surgical intervention, with a total of 193 patients. These studies used a combination of surgical techniques, predominantly unroofing, along with bypass graft and reimplantation. The perioperative results were favorable, with no operative mortalities. In follow-up, there were 4 patients with evidence of ischemia (1 unroofing patient, 3 bypass graft patients including 1 with stenosis at left internal mammary artery–left anterior descending anastomosis and 1 right internal mammary artery graft occlusion). In 1 patient, severe aortic regurgitation developed after unroofing.

Similarly, Ponzoni and coworkers[27] performed a meta-analysis of outcomes in pediatric patients and young adults (maximum age, <30 years) including 13 studies and 384 patients. The pooled results revealed an early and late mortality of 0% and 0.1%, respectively. Cardiac reoperation for all causes occurred in 3%; for aortic regurgitation, it was 0.1%. At last follow-up, 9% were symptomatic and recurrent SCA occurred in 1%; 92% were repaired with unroofing.

Independent of technique employed, all risk factors of the anomalous coronary course need to be addressed and adequacy of the repair must be checked after surgery, commonly by CTA before discharge. Evaluation for ischemia is also recommended 3 months after discharge and before resumption of sports activities.[1] Patients require ongoing surveillance for angina-related symptoms, and workup should be initiated should such symptoms arise.

MIDTERM OUTCOMES AFTER SURGICAL REPAIR

Whereas most surgical series have focused on perioperative and short-term outcomes, several studies have reported midterm results with cohorts observed for ≥5 years. Mainwaring and colleagues[19] described 115 patients with 6-year follow-up without a significant number of adverse events. Similarly, the studies of Balasubramanya and coworkers[39] of 66 patients with follow-up of 5.6 years and Doan and coworkers[46] of 52 surgical patients with a 5.5-year follow-up did not find midterm complications. Patlolla and colleagues[8] reported on 9.5-year follow-up of 148 patients with a median age of 44 years. There was not a cumulative incidence of complications during follow-up. There were 5 late deaths, 3 noncardiac and 2 unknown. Nonetheless, there are reports of SCA and ischemia after surgery (Table). For example, in the study by Sachdeva and coworkers[28] of 63 pediatric patients, 3 had SCA without a residual anatomic abnormality identified. Given that many patients are undergoing repair in their teenage years, it will be important to assess long-term outcomes. It remains to be determined whether a cumulative risk of aortic valve replacement becomes important in the long term, particularly with commissural post manipulation. Similarly, groups have reported patch aneurysm or thrombosis[35] that may become important over time. Lifelong follow-up is recommended in patients with AAOCA. The frequency of follow-up and testing performed is dependent on coronary diagnosis, age of the patient, activity level, symptoms, and whether the patient has undergone surgical repair.[1]

PERCUTANEOUS CORONARY INTERVENTIONS

Although there have been reports of percutaneous coronary stenting to relieve the proximal intramural obstruction,[42] placement of a stiff metal frame in a dynamic compressible vessel and potentially protruding into the aorta may be prone to target lesion failure. Percutaneous intervention should therefore be reserved for appropriate clinical scenarios in nonsurgical adult candidates until we have long-term results of patency, risk of fracture, and target lesion revascularization.

CONCLUSION AND REMAINING QUESTIONS

Surgical intervention remains the standard of care for patients with AAOCA who have documented ischemia, with surgery focusing on completely addressing all aspects that could contribute to coronary obstruction. Research is ongoing in modalities to better assess for inducible ischemia and to guide decision-making as well as in refining anatomic factors involved in risk stratification. Further data are required in understanding the natural history and risk of adverse events in older adults without evidence of ischemia.

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