31 Congenital Coronary Anomalies

Silvana Molossi, S. Kristen Sexson Tejtel, Prakash M. Masand, Athar M. Qureshi, Carlos M. Mery

Congenital coronary anomalies are a heterogeneous group of abnormalities with variable clinical presentation. This chapter will focus on anomalous aortic origin of a coronary artery (AAOCA) and myocardial bridges (MB). For anomalous left coronary artery from the PA (ALCAPA) see Chapter 30.

AAOCA is an abnormality of the origin or course of a coronary artery that arises from the aorta (Figure 31-1). Its prevalence is unclear, but likely between 0.2% and 0.9%. MB is a segment of a coronary artery that travels within the myocardium instead of having a normal epicardial course. The prevalence of MB is estimated to be approximately 25% and, thus many times is considered a normal variant.

An increasing number of children and adolescents are being diagnosed with AAOCA or MB following routine athletic preparticipation screening, presence of a murmur, or an abnormal ECG. Due to multiple controversies on risk stratification and incomplete understanding of the natural history of these conditions, the Coronary Anomalies Program (CAP) was developed at TCH in December 2012. Since its inception, over 250 patients have been evaluated and managed as part of the program.

Pathophysiology and Clinical Presentation

AAOCA

AAOCA is the second most common cause of sudden cardiac death (SCD), especially when the anomalous coronary originates from the opposite sinus of Valsalva and takes an *interarterial* (coronary travels between the great vessel) or *intramural* (proximal segment of the coronary travels within the wall of the aorta prior to exiting into the mediastinum) course. However, the pathophysiological mechanisms that predispose to SCD are not fully understood. It appears that age and anatomy play an important role in the development of symptoms, signs of myocardial ischemia, and/or SCD. Reports of SCD in children younger than 10 years of age are uncommon, with most events appearing to affect individuals between 10 and 30 years of age. Moreover, despite anomalous right coronary artery (ARCA) being approximately 4-6 times more prevalent than anomalous left coronary artery (ALCA), ALCA is associated with 85% of SCDs related to AAOCA and, hence, a more lethal condition than ARCA.

Occlusion and/or compression of the anomalous vessel during exercise may lead to reduced perfusion with myocardial ischemia and subsequent ventricular arrhythmia. However, it is unknown why an athlete can exercise intensely for several years with no symptoms until the sentinel event occurs. Several mechanisms have been proposed, including compression of the intramural segment of the coronary during vigorous exercise, compression of the interarterial segment between the aorta and the PA, and ostial abnormalities including an acute-angle takeoff, a slit-like ostium that may collapse with aortic expansion, or frank ostial stenosis. The intercoronary pillar (a thickening of the wall of the aorta that extends cranially from the intercoronary commissure of the aortic

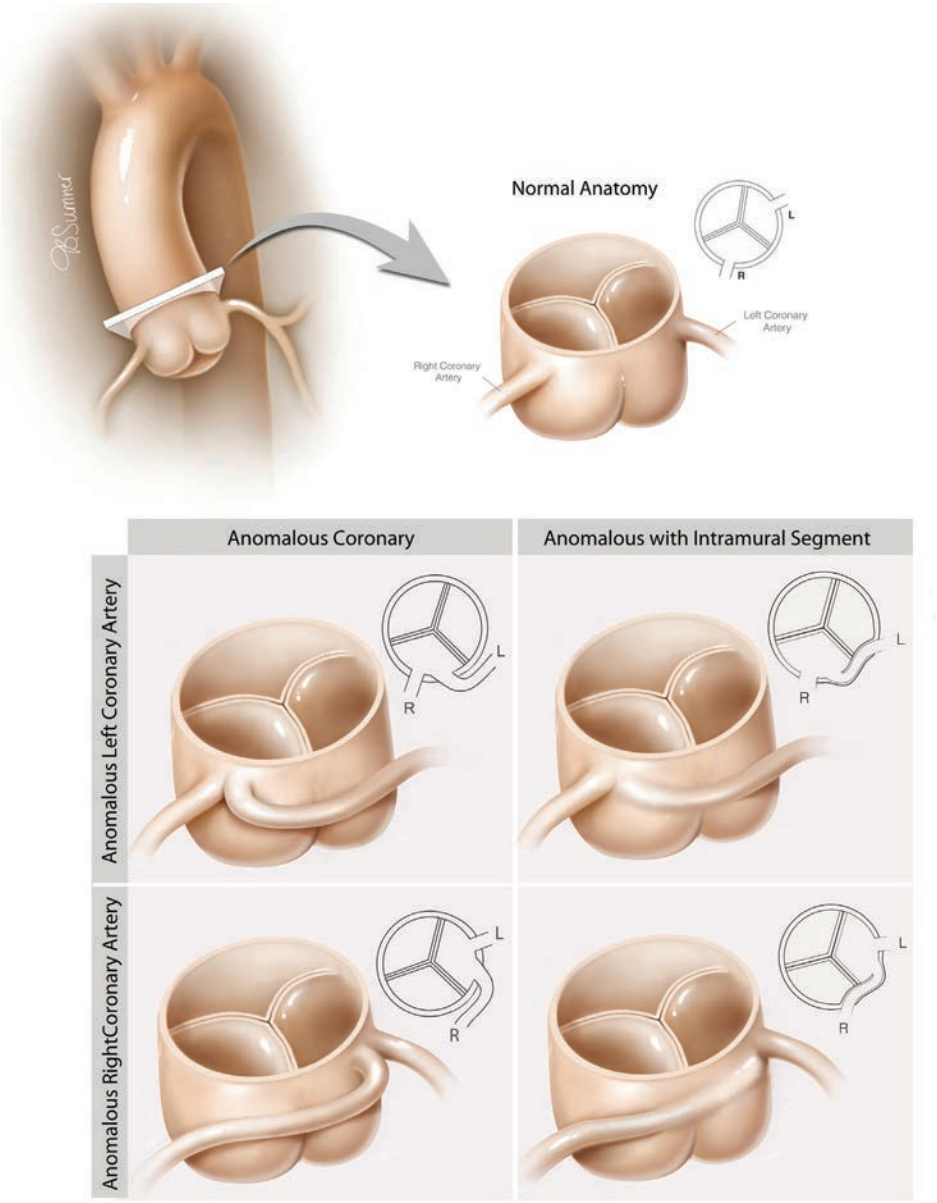
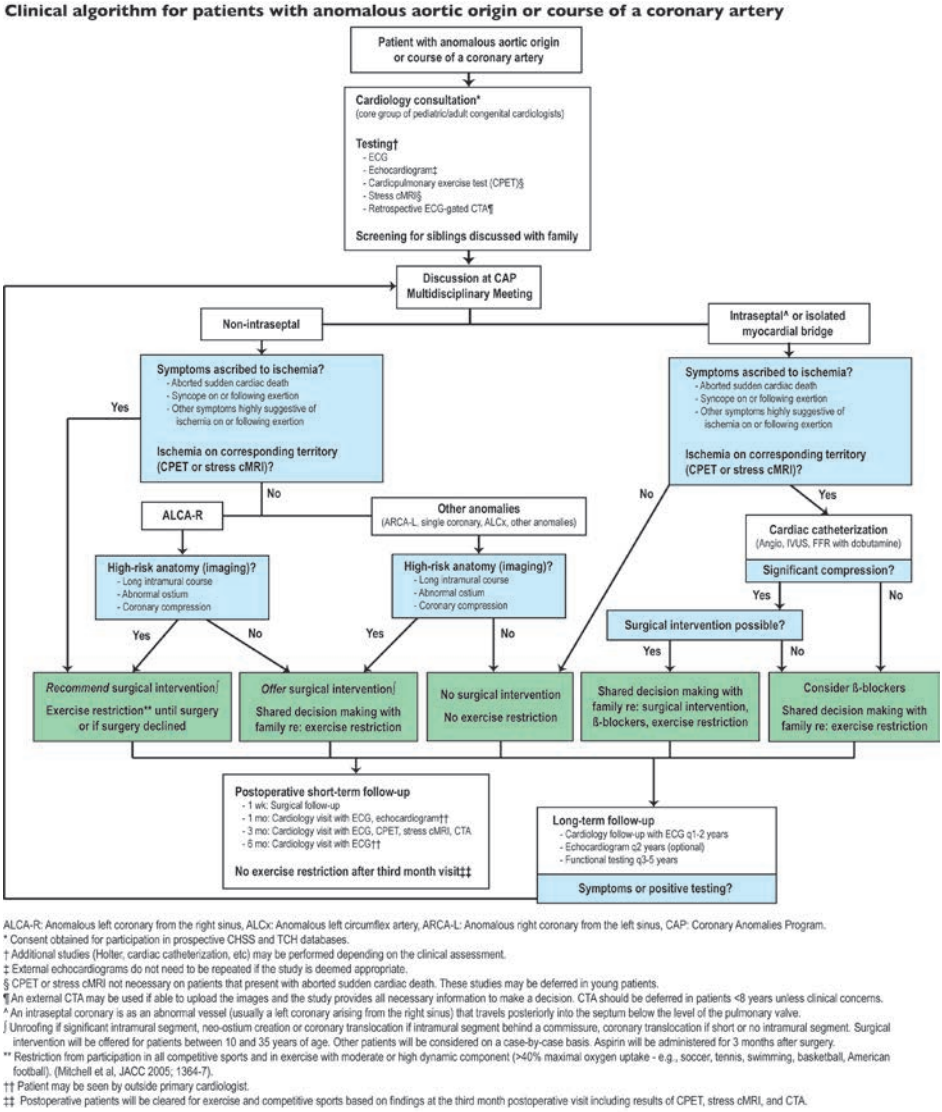


Figure 31-1. Anatomy of AAOCA with and without an intramural segment.

valve up to the sinotubular junction) may play a significant role by compressing the anomalous coronary that travels behind it, as it can be quite thick in some patients. The clinical presentation of AAOCA is variable. Symptoms are usually not present in half of the patients, and an episode of aborted SCD might be the initial event in a few.



ALCA-R: Anomalous left coronary from the right sinus, ALCx: Anomalous left circumflex artery, ARCA-L: Anomalous right coronary from the left sinus, CAP: Coronary Anomalies Program.
* Consent obtained for participation in prospective CHSS and TCH databases.
† Additional studies (Holter, cardiac catheterization, etc) may be performed depending on the clinical assessment.
‡ External echocardiograms do not need to be repeated if the study is deemed appropriate.
§ CPET or stress cMRI not necessary on patients that present with aborted sudden cardiac death. These studies may be deferred in young patients.
¶ An external CTA may be used if able to upload the images and the study provides all necessary information to make a decision. CTA should be deferred in patients <8 years unless clinical concerns.
^ An intraseptal coronary is as an abnormal vessel (usually a left coronary arising from the right sinus) that travels posteriorly into the septum below the level of the pulmonary valve.
∫ Unroofing if significant intramural segment, neo-ostium creation or coronary translocation if intramural segment behind a commissure, coronary translocation if short or no intramural segment. Surgical intervention will be offered for patients between 10 and 35 years of age. Other patients will be considered on a case-by-case basis. Aspirin will be administered for 3 months after surgery.
** Restriction from participation in all competitive sports and in exercise with moderate or high dynamic component (>40% maximal oxygen uptake - e.g., soccer, tennis, swimming, basketball, American football). (Mitchell et al, JACC 2005; 1364-7).
†† Patient may be seen by outside primary cardiologist.
‡‡ Postoperative patients will be cleared for exercise and competitive sports based on findings at the third month postoperative visit including results of CPET, stress cMRI, and CTA.

Figure 31-2. Current algorithm for diagnosis and management of patients with AAOCA and intramyocardial coronaries at TCH.

The other half of patients may present with chest pain, palpitations, shortness of breath, dizziness or syncope, during or immediately following exertion. These symptoms are quite common in the outpatient pediatric cardiology practice and this young population is the one at risk for SCD, making the validation of symptoms as it relates to the the diagnosis even more complex. Not infrequently, symptoms may be attributed to bronchospasm from asthma rather than a manifestation of myocardial ischemia. In a

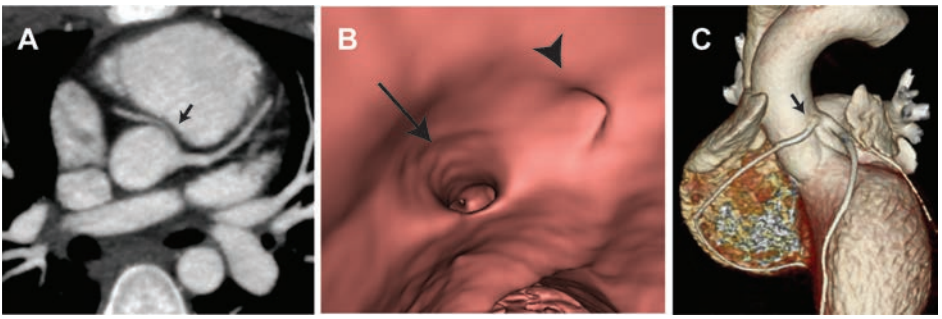


Figure 31-3. CTA in patients with AAOCA. A) Axial image in a 15-year-old demonstrating an ARCA from the left sinus, with narrow caliber of the vessel in its proximal interarterial course (arrow). B) Virtual angioscopy view showing a normal-appearing round configuration of the left main coronary artery ostium (arrow) and an elliptical, slit-like configuration of the anomalous right coronary artery ostium (arrowhead). C) Coronal volume-rendered image from a coronary CTA showing ARCA from the left sinus (arrow), with the anomalous coronary coursing just above the intercoronary commissure.

recent analysis of patients from the CAP at TCH, half of the patients were incidentally diagnosed, with another quarter presenting with symptoms on exertion, and 4 out of 163 patients presenting with aborted SCD.

MB

Although MB can be considered a normal variant, clinical manifestations vary widely. It has been suggested that myocardial bridges surrounding the coronary can compress and twist the vessel, therefore compromising flow. The functional significance may relate to the length and depth of the segment embedded in the myocardium, and the presence of more than one bridged segment.

The large majority of MB are asymptomatic. However, patients may manifest angina or angina-like symptoms, including exertional chest pain, exertional dyspnea, syncope, troponin leak, ventricular arrhythmia, myocardial infarction, and SCD.

Intraseptal Coronaries

Some patients may have a complex coronary anatomy in which an ALCA arises from the right sinus of Valsalva or from a single right coronary artery and dives into the ventricular septum within the RVOT. The coronary travels in an intramyocardial/intraseptal fashion for a variable length prior to becoming epicardial. Even though many patients with this anatomy are asymptomatic, patients can have significant coronary compression and symptomatology.

Diagnosis

Figure 31-2 shows our most recent CAP algorithm for workup and management of patients with coronary anomalies. All patients are evaluated by a specialized group

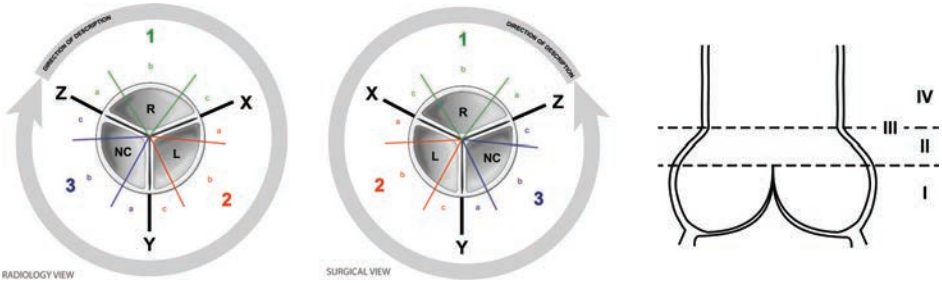


Figure 31-4. Topographic map for description of coronary ostia. The left panels describe the location of the coronary ostia based on the radiologic or surgical views. The right panel allows description of the height of the ostium (I: centrally located, II: above the aortic valve commissures but below the sinotubular junction, III: at the sinotubular junction, IV: above the sinotubular junction).

of cardiologists and undergo a standardized workup. Their data is then discussed at dedicated multidisciplinary meetings of the CAP.

- **ECG.** In the absence of ischemia, the resting ECG will be normal.
- **Echocardiogram.** May suspect AAOCA and in experienced hands, may be diagnostic. Its role is limited in MB. It is important to document ventricular function, the presence of intracardiac shunting, and other cardiac abnormalities.
- **Exercise stress test (EST).** An EST with measurement of MVO_2 is performed in all patients. Although the EST is normal in most (even those that might later present with SCD), it may disclose inducible myocardial ischemia. A positive EST is helpful for risk stratification although a normal EST does not rule out a high-risk lesion.
- **CTA (Figure 31-3).** Retrospective ECG-gated CTA provides excellent spatial resolution and is routinely used for noninvasive evaluation of coronary anatomy in children at TCH. Images are then postprocessed using a 3D workstation. Most studies are performed without the use of pharmacologic agents but beta-blockers may be needed when evaluating coronary ostial issues in patients less than 4-5 years of age. The use of new-generation scanners has significantly decreased the amount of ionizing radiation administered (approximately 2-5 mSv). The CTA report is standardized and includes information about the location of all coronaries, the presence of interarterial or intramural portions (and their length), ostial morphology and relationship, and coronary course including the relation to the intercoronary commissure or pillar. A standardized topography map is used to determine the location of the ostia (Figure 31-4).
- **Stress cardiac MRI (CMR).** Due to its excellent sensitivity and specificity to demonstrate the presence of myocardial ischemia, CMR has substituted nuclear perfusion imaging as the test of choice for functional imaging at TCH. The study is performed using dobutamine, which increases myocardial contractility while decreasing SVR, therefore mimicking exercise physiology. The perfusion sequences

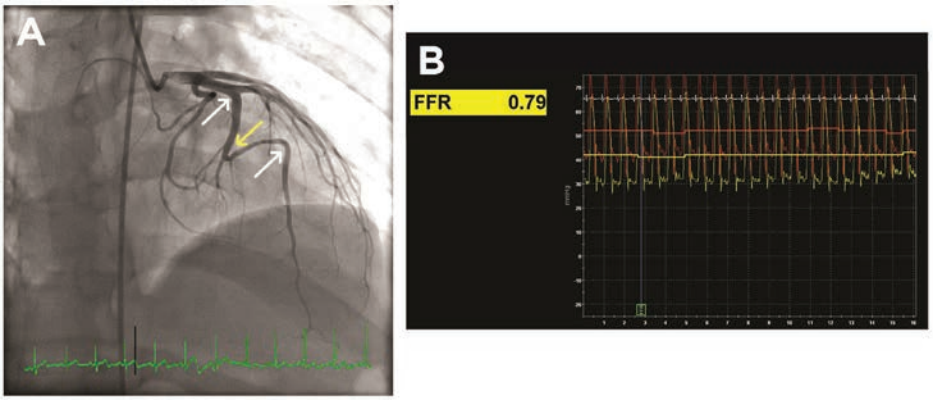


Figure 31-5. A) Left coronary artery angiogram in a 15-year-old with a long intramyocardial course of the mid left anterior descending (LAD) (corresponding to the coronary segment between white arrows) with a V- shaped hypoplastic intramyocardial segment of the LAD (yellow arrow). The fractional flow reserve (FFR) decreased from a baseline value of 0.92 to 0.79 with dobutamine infusion (B).

are performed at rest and at peak stress, with IV injection of gadolinium, to assess myocardial perfusion abnormalities and myocardial scarring.

- **Cardiac catheterization.** Cardiac catheterization may be indicated in: 1) patients in whom the coronary anatomy is not well defined with noninvasive testing, 2) patients with MB with symptoms or equivocal noninvasive testing, or 3) postoperative patients with symptoms or equivocal noninvasive testing. Coronary angiography, fractional flow reserve (FFR) testing, and intravascular ultrasound (IVUS) are performed. FFR (the ratio of pressure distal to the lesion to the pressure proximal to the lesion) is performed with the administration of intravenous adenosine (140 mcg/kg/min for 3 min) and/or dobutamine (20-40 mcg/kg/min) to achieve a heart rate of at least 75% of the predicted peak exercise heart rate. A positive FFR is considered to be <0.80 with provocative testing (Figure 31-5).

Indications / Timing for Intervention

Indications for intervention in AAOCA and MB are controversial. In general, indications depend on the type of anomaly identified and the presence of symptoms concerning for ischemia (i.e., chest pain or syncope upon or immediately following exertion), troponin leak, and/or evidence of ischemia on myocardial functional studies (i.e., EST, nuclear perfusion test [no longer used at TCH], stress CMR) (Figure 31-2). Additionally, the age of the patient will influence decision making. It is rare to entertain intervention in patients <10 years of age given the rarity of SCD, although intervention may be indicated if there is evidence of ischemia.

Due to the controversies surrounding the indications for intervention in AAOCA and the anxiety associated with this diagnosis, a long discussion with the family is of utmost importance. Patients with symptoms or evidence of ischemia are recommended

surgical intervention. Most asymptomatic patients with ARCA do not require intervention due to the low risk of SCD. However, surgical intervention may be offered for patients with significant ostial stenosis or hypoplasia, or a long intramural segment (>5 mm) with a narrowed caliber of the vessel. Due to the higher risk of SCD, intervention is usually offered to patients with ALCA from the opposite sinus. It is unclear whether patients with ALCA and no intramural segment should undergo surgical intervention; a discussion with the family is important. An anterior and prepulmonic ALCA is likely a benign variant and no intervention is required. Anomalous circumflex coronaries are also considered normal variants unless there is evidence of ischemia.

For patients with MB or intramyocardial coronaries, intervention is considered whenever there is evidence of ischemia on cardiac catheterization with FFR measurement. If surgical intervention is considered high risk, medical management with beta-blockers or calcium-channel blockers may be considered.

Strategies for intervention should always be the result of shared decision-making among all involved in the care of the patient, including the cardiologist, the surgeon, and the family. Surgical intervention may be considered or offered if there are enough concerns to impose exercise restrictions in patients with these anomalies. There are several concerns recommending exercise restriction to patients, including the difficulty of children and adolescents have adhering to such recommendation, the possibility of SCD occurring at rest or with minimal activity, the psychological and emotional consequences of restricting exercise in a child or adolescent, and the known health consequences of not exercising. As such, it is rare for exercise restriction to be recommended as a strategy at TCH.

Surgical Repair

AAOCA

Preoperative TEE is performed prior to surgical intervention to rule out the presence of an intracardiac shunt (e.g., PFO) that needs to be repaired at the time of surgery. The procedures are performed via median sternotomy and under CPB using aorto-bicaval cannulation. After cardioplegic arrest and left-heart venting through either the pulmonary vein or the atrial septum, an oblique aortotomy is performed and the coronary anatomy is inspected. Documentation of the location of the ostia (Figure 31-4) and length of intramurality is critical.

Surgical unroofing of the anomalous coronary artery is the treatment of choice at TCH for patients with a long intramural segment that travels above the level of the intercoronary commissure (Figure 31-6). The wall between the coronary artery and the aortic lumen is excised and the intimas of the aortic wall and the coronary are attached with a series of fine interrupted sutures in order to evert the edges, increase the coronary lumen, and exclude the aortic-wall fatty tissue from the circulation. By unroofing a long intramural segment, the ostium is augmented and in essence moved to the correct sinus, away from the intercoronary pillar. Unroofing of a short intramural segment may augment the size of the ostium but fail to reposition the ostium away from the intercoronary pillar, potentially causing persistent narrowing of the coronary as it travels behind the intercoronary pillar. In these cases, or in patients where the

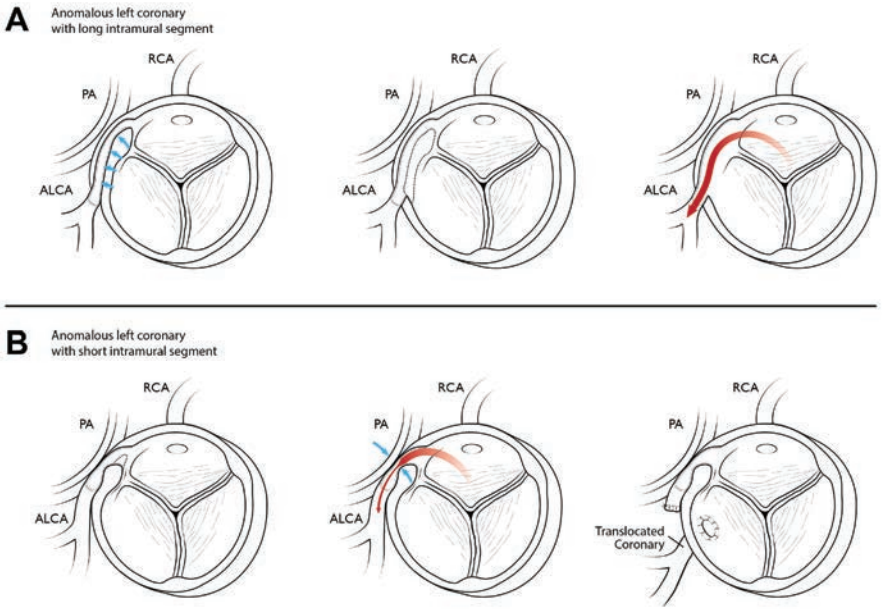


Figure 31-6. Surgical management of patients with AAOCA. A) Patient with AAOCA and a long intramural segment. By unroofing a long intramural segment, the coronary ostium is enlarged and the ostium is moved to the correct sinus and away from the thick intercoronary pillar, resulting in an unobstructed coronary ostium. B) Patient with AAOCA and a short intramural segment. Unroofing of a short segment may improve the size of the ostium but the anomalous coronary may continue to arise from the incorrect sinus and/or in close relationship with the thick intercoronary pillar, which can continue to compress the anomalous coronary. In this scenario, a coronary translocation may provide a better surgical alternative.

intramural segment travels below the level of the aortic valve, a *coronary translocation* may be a better alternative (Figure 31-6). It is important to note that different than coronary translocation in an arterial switch operation, in AAOCA, the anomalous vessel is transected as it comes out of the aortic wall (without an aortic button) and sutured circumferentially to the correct sinus. The long-term consequences of a circumferential anastomosis on a small coronary artery are unclear.

Other surgical procedures that may be used for AAOCA include the *creation of a neoostium* in the correct sinus for patients with a very long intramural segment traveling below the level of the aortic valve, and *anterior or lateral pulmonary translocation* PA (which has not been used at TCH) to theoretically prevent the compression of the interarterial segment of the coronary by the PA. Figure 31-7 shows an algorithm indicating how the optimal surgical procedure is chosen at TCH.

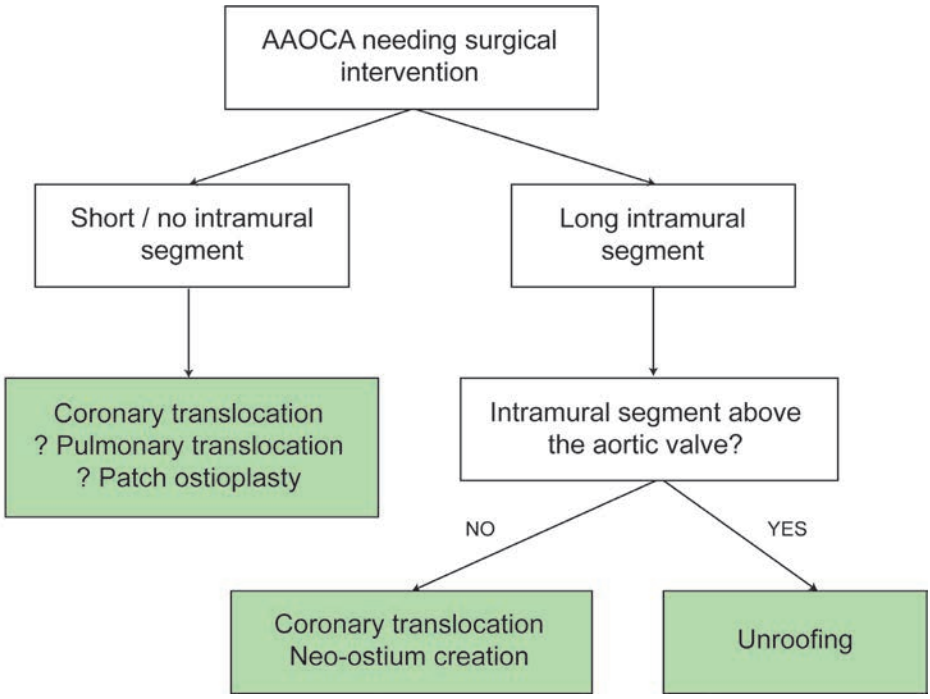


Figure 31-7. Surgical decision-making algorithm for AAOCA.

MB

The management of MBs is also performed under CPB and with cardioplegic arrest and venting of the left heart. It is important to study the anatomy of the MB and its relationship with the different coronary branches on CTA and/or cardiac catheterization to aid with intraoperative identification of the MB. Once the MB is identified, the myocardium above the coronary artery is carefully incised until the intramyocardial coronary is completely unroofed.

Intraseptal Coronaries

The surgical management of intraseptal coronaries is difficult due to their anatomy. Some proposed surgical interventions include: 1) unroofing the intramyocardial segment behind the pulmonary valve; 2) excising the pulmonary root (similar to what is performed during the Ross procedure), unroofing the myocardium above the coronary, and reimplanting the pulmonary root into the RVOT below the level of the coronary; and 3) opening the RVOT transversely, unroofing the myocardium above the coronary, and reconstructing the defect with a patch.

Postoperative Management

Patients are generally extubated in the OR and then transferred to the CICU. No inotropes are usually required for these patients. An ECG is obtained on arrival and on postoperative day 1. Approximately 70-80% of patients will have diffuse ST changes consistent with early repolarization. Classic changes involve elevation of the J point and diffuse scooping of the ST segments. As long as the changes are diffuse and consistent with early repolarization ("pericarditis"), no intervention is required. Localized ST changes, in particular related to the region of the involved coronary, should be investigated.

Patients are typically transferred to the acute floor on postoperative day 1. Low-dose aspirin is started and continued for 3 months to avoid thrombi formation in the area of surgical manipulation (tacking sutures). Patients are discharged once the chest tubes are out and discharge studies (echocardiogram and CXR) are performed, usually 4-7 days postoperatively. Patients are seen in surgical clinic 1 week after discharge, and then by the cardiologist at 1 monthwith ECG and echocardiogram. At 3 months postoperatively, patients undergo echocardiogram, ECG, EST, stress CMR, and CTA to assess the results after surgery.

Complications

- **Pericardial effusion.** The development of pericardial effusions after surgery for coronary anomalies affects approximately 10% of patients. The etiology is unclear. We have now elected to open the right pleural space and leave the pericardium open on these patients to avoid the development of significant effusions.
- **Coronary ischemia.** Ischemia is very rare but should be entertained if there are localized ECG changes or wall-motion abnormalities on echocardiography.

Long-Term Follow-Up

All patients evaluated are followed up for life. Patients are generally seen with a clinical visit and an ECG every year. For surgical patients, myocardial functional studies are performed at 3-5 years after surgery to reevaluate potential long-term effects of the surgical procedure. Long-term follow-up for patients with AAOCA is critical to eventually define the optimal management for these patients.

Suggested Readings

Agrawal H, Mery CM, Krishnamurthy R, Molossi S. Anatomic types of anomalous aortic origin of a coronary artery: A pictorial summary. Congenit Heart Dis 2017;12:603-606.

Agrawal H, Molossi S, Alam M, et al. Anomalous coronary arteries and myocardial bridges: Risk stratification in children using novel cardiac catheterization techniques. Pediatr Cardiol 2017;38:624-630.

Agrawal H, Qureshi AM, Alam M, et al. Anomalous aortic origin of a coronary artery with an intraseptal course: novel techniques in hemodynamic assessment. BMJ Case Rep 2018;pii:bcr-2018-225707.

Doan TT, Wilkinson JC, Agrawal H, et al. Instantaneous wave-free ratio (iFR) correlates with fractional flow reserve (FFR) assessment of coronary artery stenoses and myocardial bridges in children. J Invasive Cardiol 2020;32:176-179.

Doan TT, Zea-Vera R, Agrawal H, et al. Myocardial ischemia in children with anomalous aortic origin of a coronary artery with intraseptal course. Circ Cardiovasc Interv 2020; 13(3):e008375. doi: 10.1161/CIRCINTERVENTIONS.119.008375. Epub 2020 Feb 27.

Mery CM, De Leon L, Molossi S, et al. Outcomes of surgical intervention for anomalous aortic origin of a coronary artery: A large contemporary prospective cohort study. J Thorac Cardiovasc Surg 2018;155:305-319.

Mery CM, Lopez KN, Molossi S, et al. Decision analysis to define the optimal management of athletes with anomalous aortic origin of a coronary artery. J Thorac Cardiovasc Surg 2016;152:1366-1375.

Molossi S, Agrawal H. Clinical evaluation of anomalous aortic origin of a coronary artery (AAOCA). Congenit Heart Dis 2017;12:607-609.

Molossi S, Agrawal H, Mery CM, et al. Outcomes in anomalous aortic origin of a coronary artery following a prospective standardized approach. Circ Cardiovasc Interv 2020;13: e008445. doi: 10.1161/CIRCINTERVENTIONS.119.008445. Epub 2020 Feb 13.

Qureshi A, Agrawal H. Catheter-based anatomic and functional assessment in anomalous aortic origin of a coronary artery, myocardial bridges and Kawasaki disease. Congenit Heart Dis 2017;12:615-618.