Chapter 50

Congenital Coronary Anomalies

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Congenital coronary anomalies may have a significant impact on myocardial perfusion and secondary ischemia, inducing left ventricular (LV) dysfunction and sudden cardiac death. This clinical relevance underlies the necessity of an understanding of the anatomy and presentation of congenital coronary anomalies and the treatment options for these anomalies. The two primary congenital coronary anomalies, anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) and anomalous course of a coronary artery between the pulmonary artery and the aorta (ACCBPAA), are reviewed here. Two other entities associated with coronary artery anomalies—coronary artery fistulas and anomalous coronary circulation—are also reviewed in this chapter.

Normally, the two main coronary arteries arise from separate ostia within the sinuses of Valsalva. The left coronary artery (LCA) then divides into the left anterior descending artery, which traverses the anterior interventricular groove, and the left circumflex coronary artery, which courses in the left atrioventricular groove. The right coronary artery (RCA) originates anteriorly from the right aortic sinus and courses along the right atrioventricular groove, commonly giving rise to the posterior descending artery.

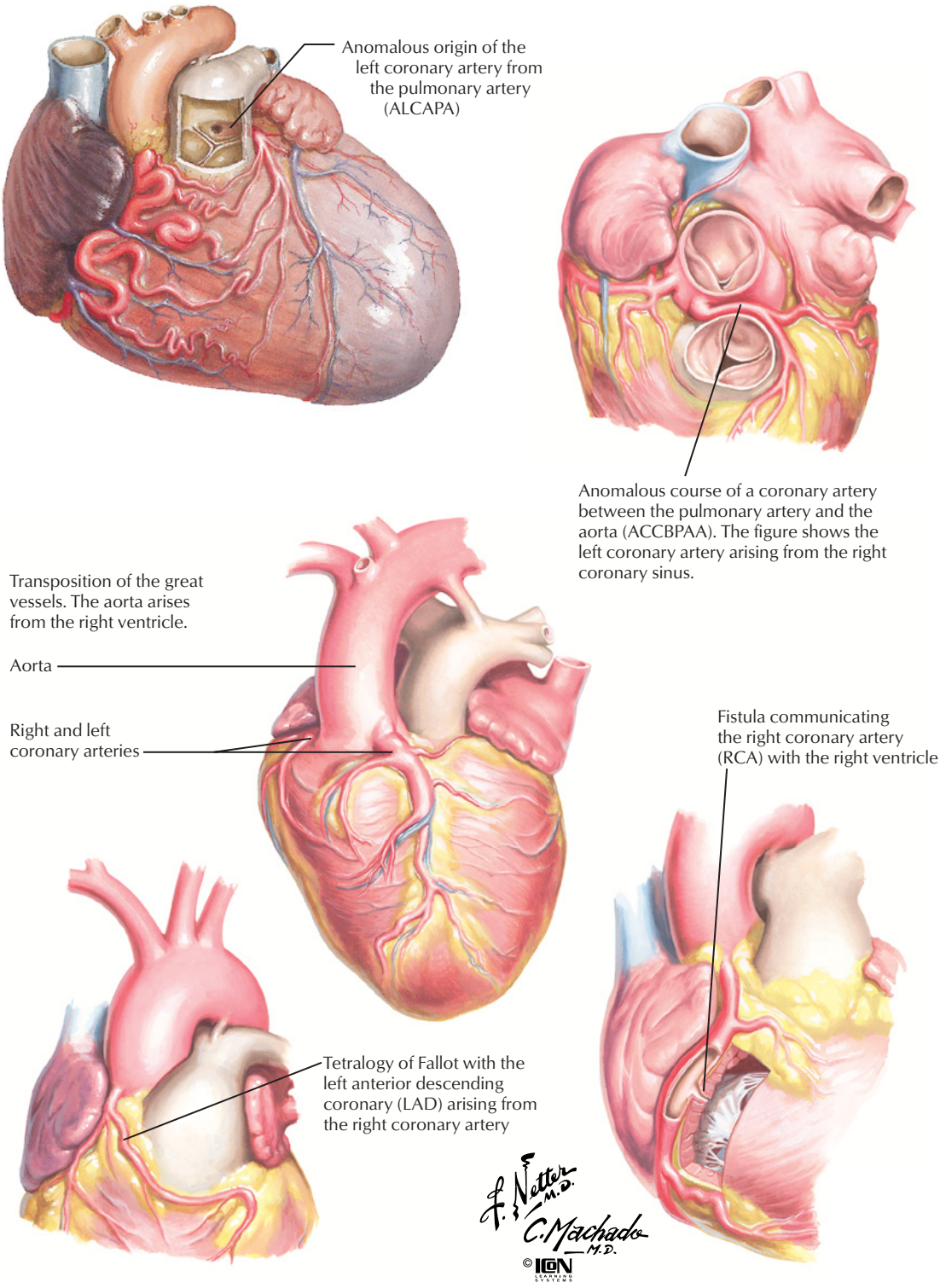
ANOMALOUS ORIGIN OF THE LEFT CORONARY ARTERY FROM THE PULMONARY ARTERY

Anomalous origin of the LCA from the pulmonary artery is a rare congenital anomaly, usually an isolated lesion, occurring in 1 in 300,000 live births (Fig. 50-1). The clinical spectrum of ALCAPA is also known as the Bland-White-Garland syndrome. Infants with myocardial ischemia typically present with failure to thrive, profuse sweating, dyspnea, pallor, and atypical chest pain upon eating or crying. Malignant arrhythmias leading to sudden cardiac death are the most extreme presentation of myocardial ischemia in ALCAPA. During the neonatal period, high pulmonary vascular resistance ensures antegrade flow from the pulmonary artery through the LCA. However, as this resistance diminishes, there is eventual reversal of flow, with left-to-right shunting through the pulmonary artery. The result is the phenomenon of "coronary steal," with LV perfusion becoming dependent on collateral circulation from the RCA.

Because infantile circulation has little or no coronary collateral development, ALCAPA leads to severe myocardial ischemia, with resultant LV dysfunction and dilation. Because of papillary muscle ischemia, and as the left ventricle dilates, mitral valve regurgitation occurs. Without surgical intervention and correction of the anomaly, patients die within weeks to months after birth. Patients who survive to adulthood, secondary to the presence and formation of collateral circulation, may remain asymptomatic despite subclinical ongoing ischemia. Arrhythmic sudden death purportedly occurs in 80 to 90% of patients by 35 years of age.

Although ALCAPA is rare, a high index of suspicion should be present for infants presenting with signs of myocardial ischemia or dysfunction. The most frequent confounding diagnosis is dilated cardiomyopathy. Both conditions may present with cardiomegaly, a murmur of mitral insufficiency, and ischemic signs on ECG. Two-dimensional echocardiography and coronary angiography typically clarify the diagnosis. Echocardiographic examination alone may be sufficient to achieve diagnosis, if this examination reveals an enlarged RCA with global hypokinesis and dilation of the left ventricle. Pulsed and color flow Doppler examination may delineate a left-to-right shunt. Two-dimensional echocardiographic evaluation may permit the visualization of the anatomic origin of the ALCAPA and the assessment of the degree of mitral insufficiency. Although not

Figure 50-1 **Congenital Coronary Artery Anomalies**



essential, coronary angiography or ventriculography, may be performed if ALCAPA is suspected but not visualized on echocardiography. Coronary angiography also assists in excluding other anatomic etiologies for ischemia and ventricular dysfunction.

Surgical correction remains the gold standard of therapy, but important changes in surgical technique have resulted in improved outcomes. Surgical repair involves direct reimplantation of the anomalous LCA into the aorta by transferring it with a button of pulmonary artery (Fig. 50-2). Variations of this technique are used when it is necessary to overcome anatomic challenges of the length and the course of the LCA for reimplantation. In adults, in whom reimplantation is more technically challenging, bypass grafting with the left internal thoracic artery is an equally effective approach.

After reestablishment of a two-coronary system, the previously dilated RCA returns to normal size, with regression of the intercoronary collateral network. Operative mortality for all surgical techniques has improved dramatically; mortality rates ranging from 75 to 80% in the early 1980s have decreased to 5 to 25%. No differences in LV function or in the late mortality rate have been demonstrated with various reimplantation or revascularization techniques, with one exception: direct ligation of the anomalous coronary was abandoned because of poor outcomes.

ANOMALOUS COURSE OF A CORONARY ARTERY BETWEEN THE PULMONARY ARTERY AND THE AORTA

Anomalous course of a coronary artery between the pulmonary artery and the aorta may result in myocardial ischemia and sudden death (Fig. 50-1). This anomaly presents with two anatomically and therapeutically distinct variations. If the RCA arises from the left aortic sinus and is nondominant, such an entity may be benign. Surgical intervention is undertaken in patients with this form of the anomaly if they have demonstrable ischemia. If the LCA arises from the right coronary sinus and courses between the aorta and the pulmonary artery, however, surgical intervention is indicated because the risk of sudden cardiac death is high in this group.

The incidence and natural history of ACCBPAA are unknown. The most significant review of this abnormality, with 242 patients, described sudden death in 59% of patients. There are no pathognomonic clinical features consistent with ACCBPAA. The diagnosis should be considered in patients with exercise-induced myocardial ischemia or sudden death. Although echocardiographic evaluation may provide valuable information, coronary angiography is essential to accurately delineate the anatomy and exclude other associated coronary disease.

Surgical options to manage this anatomic abnormality include revascularization with an internal mammary artery or a saphenous vein bypass graft or reimplantation alone. With reimplantation, a transverse aortotomy may become essential to assess the coronary ostia. When the anomalous coronary artery arises from the opposite sinus, it is necessary to detach the aortic valve commissure. The slit-like ostium, which is characteristic of ACCBPAA and partially responsible for ischemic symptoms, is opened along its longitudinal axis, and a portion of the common wall between the aorta and the coronary artery is excised, with reapproximation of the intimal surfaces. The valve commissure is subsequently resuspended with a pledgeted suture.

CORONARY ARTERY FISTULAS

Coronary artery fistulas are defined as communications with right-sided (arteriovenous fistula) or left-sided (arterio-arterial fistula) cardiac structures. The most common fistula is the RCA communicating with the right ventricle. Patients rarely present with symptoms during infancy and are frequently diagnosed in early adulthood. Often asymptomatic, a fistula is most commonly discovered during evaluation for a murmur. Echocardiographic examination may reveal evidence of a dilated or enlarged coronary artery, with color flow Doppler demonstrating the fistula. Preoperative coronary angiography ensures accurate anatomic definition for surgical planning.

Intervention prevents ventricular volume overload and resulting congestive heart failure. Although observation and transcatheter coil embolization have been described, these management options are limited to highly selected patients, because surgical treatment of coronary

artery fistulas is efficacious, reliable, and durable. If the fistula arises from the distal end of the coronary artery, ligation may be employed without cardiopulmonary bypass. Before permanent ligation, a trial occlusion of the affected coronary artery at the distal site should be performed to observe for signs of ischemia. If signs of myocardial ischemia are absent, ligation may then be performed. If the fistula arises from the midportion of a coronary artery, cardiopulmonary bypass with cardioplegic arrest allows opening of the abnormal coronary artery, where the fistula is oversewn. If coronary artery luminal compromise occurs, bypass grafting may be warranted. In other instances, the fistulous tract may be closed internally via access through the involved cardiac chamber (Fig. 50-2).

CORONARY ARTERY ANOMALIES ASSOCIATED WITH CONGENITAL HEART DISEASE

Several important forms of congenital heart disease are associated with coronary artery anomalies, which can have major implications for surgical repair. Coronary artery anomalies are particularly important in patients with tetralogy of Fallot, transposition of the great arteries, and pulmonary atresia with an intact ventricular septum (Fig. 50-1).

Coronary artery anomalies are reported in 18 to 31% of patients with tetralogy of Fallot and involve the presence of a large coronary artery crossing the right ventricular (RV) outflow tract just below the pulmonary valve. These anomalies include the origin of the left anterior descending artery from the RCA, a large conus branch across the RV outflow tract, a paired anterior descending coronary artery off the RCA, and an origin of both coronary arteries from a single left ostium. In each situation, the potential exists for damage to or severing of the coronary artery during a right ventriculotomy to correct RV outflow tract obstruction.

In pulmonary artery atresia with an intact ventricular septum, embryonic sinusoids within the right ventricle may persist and communicate with the epicardial coronary arteries in several ways. Usually this occurs in patients with diminutive RV chambers and severe RV hypertrophy. The communications may feed one or both coronary arteries and may be associated with proximal or distal coronary stenosis, or both, at the insertion site of the fistulous communications. In some patients with coronary stenosis, the coronary fistulous connections are sufficiently developed to produce an RV-dependent coronary circulation. Angiography of the RV cavity is required to demonstrate retrograde filling of one or more coronary arteries via the fistulous connection. Coronary angiography can determine whether the LV myocardium is normally perfused or whether significant segments are perfused from the right ventricle through myocardial sinusoids. In this circumstance, perfusion of parts of the left ventricle from the right ventricle must be identified before surgical repair. Decompression of the right ventricle to relieve RV outflow tract obstruction reduces RV pressure and therefore coronary artery perfusion, which can result in coronary artery ischemia and infarction during surgery.

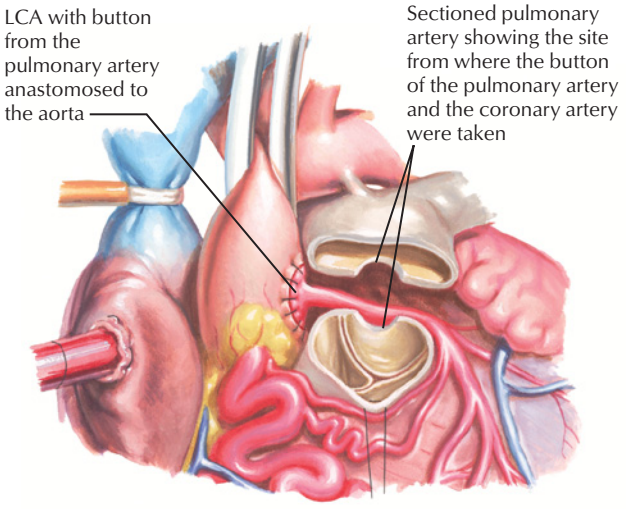
Patients who have pulmonary artery atresia with an intact ventricular septum usually require an early systemic-to-pulmonary shunt and, if the tricuspid valve and the RV chamber have growth potential, surgical relief of the pulmonary atresia. If the right ventricle is miniscule, a Fontan procedure is the definitive treatment. However, if the myocardium is perfused via the right ventricle through sinusoids because of stenotic coronary arteries, then a systemic right ventricle must be preserved as part of the Fontan operation. Cardiac transplantation may be the only option for those patients with pulmonary artery atresia with an intact ventricular septum.

The treatment for patients with a simple dextraposed-transposition (D-transposition) of the great arteries or a D-transposition of the great arteries with a ventricular septal defect is an arterial switch operation during the neonatal period (Fig. 50-2). In D-transposition of the great arteries, both in its simple form and with a ventricular septal defect, the aorta arises from the right ventricle and the pulmonary artery rises from the left ventricle. During the arterial switch procedure, the coronary arteries are transferred from the anterior semilunar valve to the posterior valve along with reversing the location of the great vessels to the appropriate ventricles. The success of the operation depends on the transfer of the coronary arteries without compromising

Figure 50-2

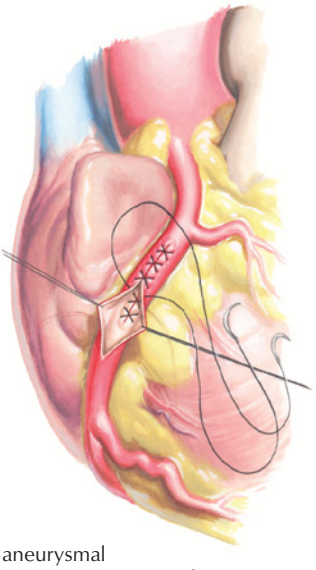
Surgical Procedures for Correction of Congenital Coronary Artery Anomalies

Surgical correction of ALCAPA



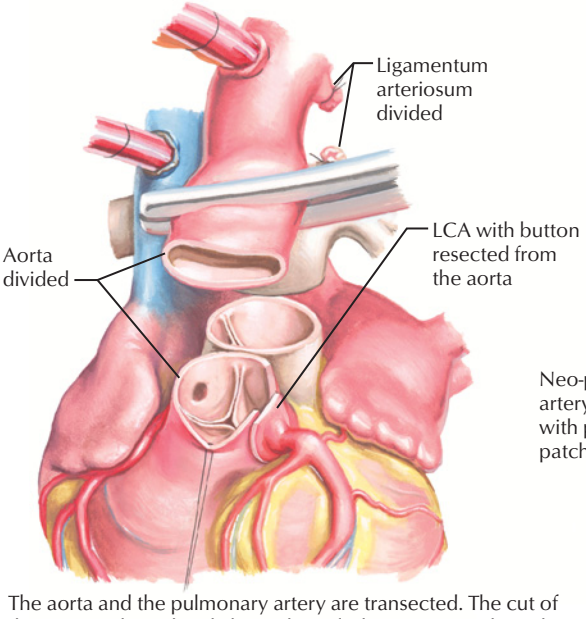
The technique involves direct reimplantation of the anomalous LCA into the aorta by transferring it with a button of pulmonary artery. Seen here: Variation with transection of the pulmonary artery.

Technique to close fistula from RCA to RV and plication of coronary aneurysm



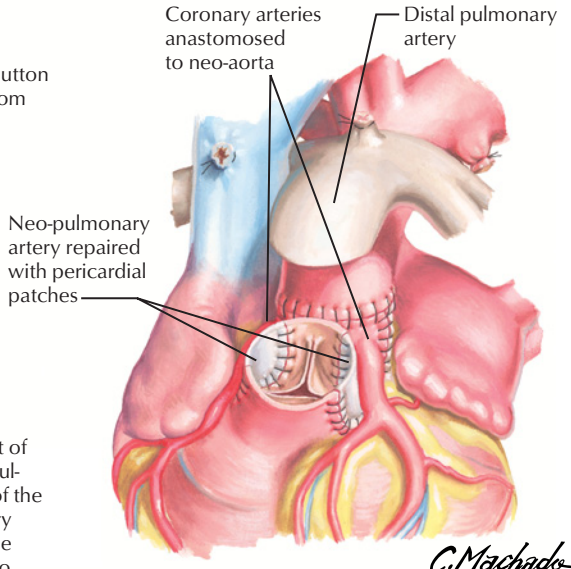
The aneurysmal coronary artery is opened and the fistula is sutured. The coronary artery is closed and the aneurysm is repaired by plication.

Arterial repair of transposition of the great arteries—First steps



The aorta and the pulmonary artery are transected. The cut of the aorta is slanted and above the Valsalva's sinuses. The pulmonary artery is divided above its valve at the same level of the transection of the aorta. Sinuses of the aorta and pulmonary artery are excised to translocate the coronary ostia from the pulmonary artery to the neo-aorta. Pericardium is utilized to reconstruct the neo-pulmonary artery sinuses.

Arterial repair of transposition of the great arteries—Last steps



the blood supply of the coronary circulation. Seven different coronary artery patterns are recognized in patients with a D-transposition of the great arteries, but normal anatomy is usually present. Although certain unusual coronary artery patterns were previously associated with an increased mortality rate, the specific coronary artery anatomy has become less important as surgical experience with this operation has improved technical approaches and overall outcomes. The presence of an intramural coronary artery, a segment of coronary artery that courses within the wall of the aorta without a separate layer of adventitial tissue between the coronary artery and the aorta, remains a difficult challenge. Although follow-up angiography after the arterial switch operation shows varying coronary artery abnormalities in approximately 10% of patients, most patients are asymptomatic.

FUTURE DIRECTIONS

Several issues of anomalous coronary arteries remain to be explored, including but not limited to the choice of the best noninvasive diagnostic imaging technique, the further pathophysiologic characterization of myocardial perfusion in patients with anomalous coronary arteries, and the definition of the indications for percutaneous intervention in adults who have symptomatic coronary disease in anomalous coronary vessels.

The tools for noninvasive imaging of anomalous coronary arteries include 16-slice multidetector spiral CT and free-breathing, three-dimensional coronary magnetic resonance angiography. Spiral CT, a noninvasive imaging modality, has comparable resolution to magnetic resonance angiography and is faster and less costly. Free-breathing, three-dimensional coronary magnetic resonance angiography is limited by availability, time, cost, and patient comfort. Magnetic resonance angiography studies are challenging to perform because of the enclosed space in which patients must be placed and the length of time to complete an evaluation. Ultimately, the method best suited for defining anomalous coronary vessels will depend on the degree of resolution offered by the technique and other considerations including cost and availability. As imaging techniques improve, noninvasive imaging for anomalous coronary arteries will likely become the standard of care. Rapid advances in imaging technologies have occurred in the last decade and promise to further improve imaging of anomalous coronary circulation in the future.

Further investigation is warranted into regional myocardial flow reserve in survivors of ALCAPA and its underlying pathology (i.e., endocardial and subendocardial fibrosis, damage to the papillary muscles, patchy myocardial necrosis, dilation of the ventricle, mitral incompetence, LCA hypoplasia of the media, distal stenosis and hypoplasia of the RCA). The physiologic issues need further definition in relation to myocardial perfusion after treatment in long-term survivors of this often lethal condition.

Anomalous coronary arteries have a reported frequency of approximately 0.64% in nonselected patients undergoing coronary angiography; it can therefore be predicted that adults who have anomalous coronary arteries will present with symptomatic coronary artery disease in these vessels later in life. Because this anatomy may offer unique challenges for interventional cardiologists, specific indications for percutaneous intervention remain to be defined in this area of improving interventional technology.

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