Management of the Adult with Arterial Switch

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ABSTRACT: Dextro-transposition of the great arteries (d-TGA) is a lethal congenital heart defect in which the great arteries—the pulmonary artery and aorta—are transposed to create ventriculoarterial discordance. Corrective surgical interventions have resulted in significant improvements in morbidity and mortality for this once-fatal congenital heart defect.

The initial palliative surgery for d-TGA was the atrial switch operation, which provided physiological correction. The Mustard and Senning "atrial switch" procedures, in which an atrial baffle is created to produce a discordant atrioventricular connection on the existing discordant ventriculoarterial connection, showed preliminary success for the correction of d-TGA. However, follow-up evaluations demonstrated increasing complications from the right ventricle utilized as a systemic ventricle, resulting in progressive right ventricular dysfunction. Thus, the search continued for an anatomical correction of d-TGA to return the great arteries to their normal ventricular connections.

The arterial switch operation (ASO), though attempted and theorized by many, was first successfully performed by Dr. Jatene and colleagues in 1975. For ASO, the distal main pulmonary artery and the distal ascending aorta are transected and then anastomosed to their respective ventricles with relocation of the coronary arteries to the neoaorta. The ASO has replaced the atrial switch operation since the 1980s and is now the standard surgical correction for d-TGA.

As more patients who have undergone ASO are living into adulthood, late complications of this procedure have become more evident. The most common late postoperative complications include coronary artery stenosis, neoaortic root dilation, neoaortic insufficiency, and neopulmonic stenosis. Adults who have undergone ASO in childhood will need follow-up with surveillance imaging and evaluation of new symptoms or declining function to prevent and manage late postoperative complications. This review describes the management strategies for common late complications in patients who have undergone ASO.

BACKGROUND

Dextro-transposition of the great arteries (d-TGA) describes the congenital heart defect in which the great arteries are transposed: The aorta arises from the right ventricle (RV) and the pulmonary artery arises from the left ventricle (LV). This discordant ventriculoarterial connection results in pulmonary and systemic circulations that run in parallel as opposed to series, with systemic venous blood flowing to the aorta and the pulmonary venous blood to the pulmonary artery.[1] The reported incidence of d-TGA ranges from 20 to 30.5 per 100,000 live births, with a strong male preponderance. It accounts for 5% to 7% of all patients born with complex congenital heart defects.[1,2] Unfortunately, without surgical correction, preoperative mortality in the neonate is approximately 30% within the first week of life and up to 90% within the first year.[1] Nevertheless, with aggressive surgical and medical interventions in the neonatal period, survival is greater than 90% with the expectation of a vigorous and robust adolescent and adult life.[1-3]

Perioperative mortality of d-TGA has improved considerably since the introduction of surgical correction. The initial palliative surgery for d-TGA was the atrial switch operation, which provided physiological correction. The atrial switch operation was introduced in the late 1950s by Dr. Ake Senning of Sweden followed shortly by Dr. William Mustard, who described a similar procedure. In the Senning atrial switch operation, atrial tissue is used to create a baffle that redirects systemic venous blood to the morphological LV and then to the pulmonary circulation. Similarly, the pulmonary venous blood flow is directed to the morphological RV, which serves as the systemic ventricle. In 1963, Dr. William Mustard described a comparable procedure in which most of the atrial septum is excised, and the atrial baffle, made from pericardial tissue or synthetic material, directs venous return. The repair imposes discordant atrioventricular connection on the existing discordant ventriculoarterial connection to create double discordance.[1,2] Therefore, with the Senning and Mustard atrial switch operations, the anatomical LV becomes the pulmonic ventricle and the RV becomes the systemic ventricle. Initially, the Mustard and Senning atrial switch procedures received widespread acceptance and were used for more than three decades. However, since the RV functions as the systemic ventricle, the RV is predisposed to

dysfunction and ultimate failure. Mid- and late-term follow-up evaluations demonstrated progressive late RV dysfunction and severe tricuspid valve regurgitation as the major complications of this procedure. Other complications include dysrhythmia, baffle obstruction, pulmonary hypertension, and even sudden death.[4,5] The high incidence of late-term complications of this procedure fueled the continued search for anatomical correction of d-TGA to return the great arteries to their normal ventricular connections. Since the mid-1950s, several attempts for anatomical correction of d-TGA were made; still, it was not clinically achieved until 1975, when Dr. Adib Dominos Jatene and colleagues first successfully performed the arterial switch operation (ASO). Mortality for the ASO remained high for several years following its introduction. However, as refinements in surgical technique, perioperative medical management, coronary translocation and the LeCompte maneuver improved in the 1980s, survival also improved.[3] Ultimately, ASO replaced the atrial switch operation and has become the palliative procedure of choice for the correction of d-TGA, with excellent long-term survival.[1,2,4]

During ASO, the great arteries are transected and returned to their normal ventricular connections. The most common technique to accomplish this is the LeCompte maneuver, in which the distal main pulmonary artery with branch pulmonary arteries are brought forward and the distal aorta is moved posteriorly. The distal main pulmonary artery is anastomosed to the proximal aorta and thus the RV. The distal ascending aorta is anastomosed to the proximal main pulmonary artery and thus connected to the LV outflow tract. Additionally, the left and right coronary artery ostia are excised from the native aortic root with surrounding aortic walls, which serve as "buttons," and moved to the neoaortic root.[1,2,4]

Though the long-term results of this operation remain unknown, several case series have described excellent long-term survival, up to 30 years, after ASO.[4,5] Case series have also revealed late complications—including coronary artery stenosis, progressive neoaortic dilation, neoaortic insufficiency, neopulmonic insufficiency, supravalvar aortic stenosis, and RV outflow tract obstruction—that can contribute to morbidity, mortality, and need for reintervention in adulthood among those who have undergone ASO.[5-7] Adults who have undergone ASO need follow-up, surveillance, and management of the long-term postoperative complications associated with this procedure.

CORONARY ARTERY OBSTRUCTION

Although ASO offers excellent long-term outcomes, coronary complications have been an important cause of both early and late mortality.[6] Jatene commented about the complexity of ASO by stating, "To divide, contrapose and re-anastomose the great arteries is not a surgical problem. The major technical difficulty [of ASO] has been the transfer of coronary arteries."[4] The degree of difficulty associated with the transfer and reimplantation of the coronary arteries is variable and depends on the specific anatomy of the origins and the course of the coronary branches.[8] The anatomy of the coronary arteries in d-TGA may vary since coronary arteries may arise from varying locations on the aortic sinuses; this is taken into account in ASO with the reimplantation of the coronary buttons to the neoaorta.[5,7,8] The prevalence of coronary artery obstruction shortly after ASO is approximately 5% to 8%, and it is a leading cause of death in this patient population.[5,8]

Most often, coronary artery events arise in the first few years following intervention, but there is limited experience with adults into the third and fourth decade of life. A long-term study found that freedom from coronary events was 88.1 ± 6.4% at 22 years.[2] One study classified late coronary complications as coronary occlusion, major stenosis (> 50% stenosis of the diameter), minor stenosis (< 50% stenosis of the diameter), and stretching of the coronary arteries causing diffuse narrowing of the coronary arteries.[8] Late coronary events may be attributed to intimal thickening and stretching as patients age. Furthermore, it has been theorized that twisting may occur at the coronary ostia, especially during periods of rapid development as the patient grows from childhood to adolescence into adulthood. Additionally, surgical coronary denervation may lead to the absence of pain perception. Therefore, the patient population that undergoes ASO can be asymptomatic and have coronary artery obstruction without typical signs of ischemic disease; ischemic disease may also be missed with routine cardiac ischemic evaluation.[5,8]

The 2018 American Heart Association (AHA)/American College of Cardiology (ACC) Guideline for the Management of Adults with Congenital Heart Disease recommends anatomic evaluation of coronary arteries with computed tomography, magnetic resonance angiography, or catheter angiography at least once in asymptomatic patients to evaluate the patency of the coronary arteries.[7] The AHA/ACC recommend further evaluation if symptoms of coronary ischemia are present.[5] In asymptomatic patients, the European Society of Cardiology (ESC) guidelines recommend a one-time evaluation of coronary vessel anatomy by cardiac catheterization and coronary angiography.[9] In the presence of myocardial ischemia, stenting or surgery may be performed for coronary artery lesions. Coronary ostial stenosis late after arterial switch may be repaired by coronary button revision, osteoplasty, interposition grafts, or coronary artery bypass surgery. It is imperative

that coronary vascularization in adults who have undergone arterial switch be performed in close collaboration with surgeons or interventional cardiologists who are experienced in revascularization for adult congenital heart disease to ensure that the anatomy of coronary and great arteries is well understood.[7]

The impact of surgical manipulation of coronary buttons on the development of coronary artery atherosclerosis is largely unknown.[8] Acquired heart disease is a risk for all patients who have undergone ASO, and all patients should have a routine cardiovascular risk assessment based on family history for premature coronary artery disease.[5,7]

NEOAORTIC DILATION AND INSUFFICIENCY

The most frequent long-term complication of ASO involves the neoaorta. In ASO, the native pulmonary root and valve functions as the neoaortic root and valve; it is theorized that the excessive pressure on the pulmonic valve in the systemic position can lead to neoaortic dysfunction, including neoaortic root dilation and neoaortic insufficiency.[6] Several studies have reported increased frequency of progressive neoaortic dilation in patients who did not have complete palliation as neonates, presence of a ventricular septal defect, or a double outlet RV. The presence of a subpulmonary ventricular septal defect may result in a relatively dilated native pulmonary annulus and root before surgical repair due to increased pulmonary blood flow.[10]

Studies have shown approximately two-thirds of patients after ASO have neoaortic root dilation. McMahon and colleagues demonstrated significant increase in size of the neoaortic root as a long-term complication among patients who underwent ASO. Fifty percent of patients in their study had moderate-to-severe neoaortic enlargement, with Z scores > 3.[11] Another study by Co-Vu and colleagues showed progressive dilation of the neoaortic root disproportionate to somatic growth; neoaortic root Z scores increased at a mean rate of 0.08 per year and may continue to increase at least 15 years after ASO without evidence of stabilization of root dimensions postoperatively.[10]

Furthermore, the presence of neoaortic dilation may have long-term implications for valve function. Progressive neoaortic root dilation may result in stress and strain on the neoaortic valve leaflets and reduced leaflet coaptation. Co-Vu and colleagues suggested an association between neoaortic root dilation and valvar dysfunction because the presence of neoaortic dilation was an important predictor for ≥ moderate neoaortic valve regurgitation. Also, the risk for neoaortic valve regurgitation appeared to increase with length of time after the operation.[10,11]

In contrast, neoaortic stenosis is seldom observed as a late-term complication after ASO.

The baseline degree of neoaortic insufficiency or supravalvar aortic stenosis should guide active surveillance intervals. Although some degree of neoaortic valve insufficiency is a common late complication of an ASO, intervention is rarely required.[7,9,10] Nonetheless, though the incidence is fairly low, neoaortic valve repair accounts for a large percent of all interventions in patients after ASO. Therefore, baseline and serial imaging with echocardiography or cardiovascular magnetic resonance should be performed every 1 to 2 years for patients with underlying valvular dysfunction.[7,9,10]

Based on the 2014 AHA/ACC Guideline for Management of Valvular Heart Disease, LV dilation and symptomology should guide neoaortic valve repair and replacement.[12] The rate of intervention on the neoaortic valve beyond 40 years of age is yet to be determined.

Although patients who have undergone ASO frequently have an increased incidence of neoaortic dilation, it is unlikely for it to progress to neoaortic root dissection.[5,10] The incidence of aortic dissection is similar to other conotruncal abnormalities. Dilation of the aortic root at dimensions < 5.5 cm is not associated with dissection and therefore is not an indication for reoperation at smaller dimensions.[10] Nevertheless, patients will need continued surveillance. For patients who have baseline neoaortic dilation, the 2018 ACC/AHA guideline recommends baseline and serial imaging with echocardiography or cardiovascular magnetic resonance.[7] Neoaortic root surgery should be considered when the neoaortic root is > 55 mm in an average adult.[7] Neoaortic root and valve dilation may modify both exercise prescription and the degree of assistance during pregnancy.

RIGHT VENTRICULAR OUTFLOW TRACT OBSTRUCTION

Late complications of obstruction at various sites through the RV outflow tract (RVOT) are the most frequent reason for reoperation and reintervention with ASO.[5,7,13] Late pulmonary stenosis occurs in 5% to 15% of patients who have undergone ASO and may occur at any level of the pulmonary tree, including at the pulmonary valve, main pulmonary artery, and branch pulmonary arteries.[1,5] RVOT obstruction occurs in sufficient severity to require reintervention in up to 10% of cases.[13]

The ASO procedure with the LeCompte maneuver orients the pulmonary artery branching just anterior to the ascending aorta and drapes the branches of the pulmonary artery around the often dilated aorta, which can lead to main and branch pulmonary artery compression, stretching, and

stenosis. Suprapulmonic stenosis secondary to stenosis at the anastomotic site can also occur as a late complication.[5] Neopulmonic insufficiency is infrequently observed, which may be due to lower stress in the pulmonary circulatory system.

Per the ACC/AHA and ESC guidelines, baseline and serial imaging with echocardiography or cardiovascular magnetic resonance should be performed to evaluate for the presence and severity of main pulmonary artery or branch pulmonary artery stenosis.[7] Right ventricular function should also be evaluated.[7,9] In the setting of inconclusive noninvasive assessment, cardiac catheterization can be used for further assessment of RVOT obstruction.[7,9,13] Symptomology and severity of stenosis should guide interventional modalities, which include balloon angioplasty, stenting, or surgery. According to the literature, the frequency of these interventions required after undergoing an ASO with the LeCompte maneuver is center specific and varies from 3% to 28%.[5] Severe RVOT obstruction not amenable to percutaneous treatment is an indication for reoperation. Pulmonary valve repair or replacement is considered when severe pulmonary valve stenosis is present and there is significant RV dilation or dysfunction.[7,9,10] Based on the AHA/ACC guideline, surgical repair of RVOT obstruction should be considered in symptomatic patients with RV systolic pressure (RVSP) > 60 mm Hg or asymptomatic patients with RVSP > 80 mm Hg.[7] Similarly, ESC guidelines support surgical repair in symptomatic patients with RVOT obstruction and RVSP > 60 mm Hg or asymptomatic patients with RV dysfunction.[9]

CONCLUSION

Excellent long-term outcomes with survival to 30-plus years after an ASO have been reported. However, with improved survival, late postoperative complications have become more apparent. The most common late complications include RVOT obstruction, neoaortic dilation, neoaortic insufficiency, and coronary artery stenosis. Other less-frequent complications that were not discussed in this review include arrhythmias and LV dysfunction. Adults who have undergone an ASO will need close follow-up surveillance and evaluation of new symptoms or declining function to adequately manage late postoperative complications.

Conflict of Interest Disclosure:
The authors have completed and submitted the *Methodist DeBakey Cardiovascular Journal* Conflict of Interest Statement and none were reported.

Keywords:
arterial switch, congenital heart disease, dextro-transposition of the great arteries, d-TGA

KEY POINTS

- Survival for dextro-transposition of the great arteries is greater than 90% with aggressive surgical and medical interventions in the neonatal period, with excellent long-term outcomes.
- Common late complications of an arterial switch operation (ASO) include right ventricular outflow tract obstruction, neoaortic dilation, neoaortic insufficiency, and coronary artery stenosis.
- Baseline and serial imaging with echocardiography or cardiovascular magnetic resonance will be needed to evaluate for postoperative late complications. Cardiac catheterization, especially coronary vascularization, in adults who have undergone arterial switch should be performed in close collaboration with surgeons or interventional cardiologists who are experienced in revascularization for adult congenital heart disease to ensure that the anatomy of coronary and great arteries is well understood.
- Patients who have undergone ASO will need continued surveillance to adequately manage late postoperative complications.

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