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Update on the Management of Adults With Arterial Switch Procedure for Transposition of the Great Arteries

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Opinion statement

The arterial switch operation (ASO) is now the most frequently performed surgical correction in individuals with dextro-transposition of the great arteries (D-TGA). Patients who undergo this procedure as neonates have overall good clinical outcomes yet continued clinical follow-up is important to evaluate for postoperative complications. In this group, the highest mortality is in the immediate postoperative period and is generally associated with reimplantation of the coronary arteries. As these patients live into adulthood, longitudinal follow-up for other ASO complications including neo-pulmonary stenosis, right ventricular outflow tract (RVOT) obstruction, or neo-aortic root dilation and resulting aortic insufficiency should be performed. In adults, extra care should be taken to identify and treat traditional cardiovascular risk factors as individuals with coronary obstruction may not present with typical anginal symptoms. Management of these patients should be performed in collaboration with an adult congenital heart center of excellence. This population offers a unique opportunity to provide timely feedback to adult congenital heart community of providers regarding late outcomes from surgical intervention and in the next decade will hopefully demonstrate a model for clinical feedback cycles in lifelong congenital care.

Introduction

Dextro-transposition of the great arteries (D-TGA) occurs in approximately 0.2 in 1000 births [1]. The most recent surgical correction for D-TGA was originally described in 1975 by Jatene and colleagues and the surgery still carries his name [2]. Surgical correction as a neonate is preferred and delay was found to be associated with an increased mortality and overall increased cost to the healthcare system [3]. As these patients are living longer and into adulthood, we will review the currently available data regarding late complications, management, and treatment after the arterial switch operation (ASO).

Prognosis and post-procedural complications

In most high-volume congenital centers, the early operative mortality associated with the ASO is approximately 3% [4]. Overall, there are good long-term (>10 years) survival rates reported as 88–97% and freedom from reoperation of approximately 82% [5, 6]. As patients progress into their adult years, postoperative late complications of the Jatene operation include the following: coronary artery obstruction, progressive neo-aortic dilatation, neo-pulmonary stenosis (supravalvular and branch pulmonary stenosis), and right ventricular outflow tract (RVOT) obstruction.

In patients who have had an ASO, the electrocardiogram may show a right bundle branch block or a first-degree AV block. Although atrial arrhythmias may occur [7], they occur less often than in patients who underwent Mustard or Senning procedures [8] where atrial or ventricular tachyarrhythmias occur around surgical sites and prosthetic material [9]. Arrhythmias in the immediate postoperative period should raise concern for ischemia related to coronary artery reimplantation and correlate with increased morbidity and mortality [10, 11]. Individuals who develop late-onset atrial arrhythmias or cardiac dysfunction, new-onset aortic insufficiency may be the inciting cause and should be excluded.

Asymptomatic coronary artery obstruction is a significant challenge in the ASO population. Coronary artery obstruction may be underdiagnosed in this population due to the lack of ischemic changes on testing [12]. The prevalence of coronary artery obstruction after ASO is approximately 8% [13–15] with coronary artery obstruction leading to the primary cause of death after ASO [16]. The coronary arteries associated with D-TGA may arise from varying locations on the aortic sinuses, and this anatomy is taken into consideration when the transfer of the coronary ostia is performed [17, 18]. During coronary artery transfer, anatomic torsion or extrinsic compression of the coronary arteries may cause ischemia. Late coronary events may also be attributed to progressive intimal thickening or stretching as patients age [13]. Residual cardiac sympathetic denervation [19] may lead to the lack of pain sensation during ischemic evaluation.

Intervention on the aortic valve after ASO has thus far rarely been needed; however, the rates of intervention on the aortic valve as this population further ages has yet to be determined. Aortic insufficiency (AI) may develop and patients should undergo clinical and imaging surveillance. The amount of AI at the time of discharge from ASO is associated with the progression to greater than moderate aortic insufficiency. One study found that 29% of patients discharged with mild AI

progressed to greater than moderate AI, whereas of those with no AI at discharge only 3.4% progressed [20]. Baseline degree of AI may therefore guide active surveillance intervals.

Individuals with D-TGA may have the concomitant presence of a ventricular septal defect (VSD) at birth. These individuals may have undergone pulmonary artery (PA) banding in order to prepare or "train" the left ventricle to be able to handle the afterload required to be the systemic ventricle [21]. The presence of a VSD, history of PA banding, or older age at the time of ASO contributes to the incidence of AI as well as aortic root enlargement [22, 23]. Fortunately, although the aortic root may dilate, patients are unlikely to have an aortic dissection [24].

Another possible sequela from ASO, RVOT obstruction, is the most frequent reason for reoperation [6, 25]. RVOT obstruction can occur after ASO at the main PA, branch PAs, pulmonic valve (PV), or the subpulmonic valve area. For supravalvular and branch PA stenosis, balloon angioplasty, stenting, or surgical intervention may be warranted. Suprapulmonic stenosis is the most common complication seen after ASO and may reflect anastomotic site or old PA band site narrowing [26]. Suprapulmonic valve stenosis and branch PA stenosis may also occur due to the Lecompte maneuver used during the ASO procedure which orients the pulmonary artery bifurcation anterior to the ascending aorta and drapes the PA branches around the often dilated aorta [27, 28] (see Fig. 1). The frequency of PA intervention reported in the literature varies between 3 and 28% and is often center specific [4, 26, 29]. Multidisciplinary review at an adult congenital heart disease (ACHD) center with ACHD cardiologists, interventionalists, surgeons, and imagers is extraordinarily useful in determining indications, timing, and methods of repair as the guidelines continue to evolve.

Management and recommendations

All patients with D-TGA should be seen and followed annually for development of symptoms or complications at an ACHD center of excellence. As this

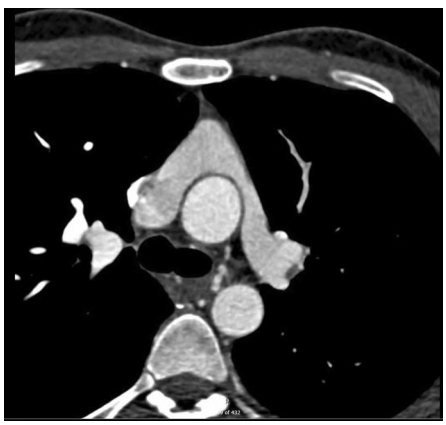

Fig. 1. Coronary CTA image of a patient with a history of D-TGA and ASO in which a Lecompte technique was used. The resulting PA bifurcation anterior to the ascending aorta in the chest is shown.

population is still young, there is a paucity of data regarding management of ASO patients and the frequency of surveillance. The 2008 ACC/AHA guidelines [30] recommend that noninvasive testing be performed every 3–5 years to evaluate for ischemia. After ASO repair, at least one study to evaluate patency of the coronary arteries either invasively or noninvasively is recommended. The ESC guidelines [31] recommend a one-time invasive assessment of coronary vessel anatomy by cardiac catheterization be performed in asymptomatic patients; however, the risk/benefit of invasive assessment, whether singular or several, needs to be considered carefully for each individual patient. At our institution, the use of coronary computed tomography angiogram (CTA) has significantly decreased the need for invasive testing, although serial testing with this modality may again carry some risk and warrants further discussion. Continued follow-up with echocardiography is recommended in patients with previous ASO at least every 2 years at a center with experience in adult congenital echocardiography [30].

Current guidelines recommend that in the ASO population, long-term complications requiring intervention may include stenting or surgery for coronary arteries that are causing ischemia and/or surgical repair of RVOT obstruction in symptomatic patients with right ventricular (RV) systolic pressures >60 mmHg or asymptomatic patients with RV systolic pressures >80 mmHg. The guidelines also support surgical RVOT obstruction repair when RV dysfunction develops regardless of symptoms [31].

Although some patients after ASO were found to have impairment of chronotropic response, it does not usually interfere with exercise capacity [24, 32]. Physical activity leads to a higher quality of life in D-TGA patients after ASO [33] and, as in other ACHD lesions, should be prescribed. In the setting of a normal stress test, patients can participate in all sports if there is normal ventricular function and no documentation of an arrhythmia. If ventricular dysfunction or mild hemodynamic abnormalities are found, low to moderate static/dynamic exercise may be evaluated on an individual basis [34]. In the presence of aortic dilatation, although the guidelines do not specifically address that issue, clinicians may consider recommending aerobic over isometric exercise.

As the patients reach childbearing age, pre-conception counseling is recommended at an ACHD center in collaboration with maternal fetal medicine. The hemodynamic changes associated with pregnancy can present risk to the patient [35] and a pre- and postpartum plan should be discussed. Evaluation for ischemia, valvular dysfunction, arrhythmia, and aortic dilatation should be included in the preconception visit to guide risk stratification [36].

Conclusion

Individuals with D-TGA who undergo an ASO a young age thus far have excellent long-term outcomes. As these patients age, traditional cardiovascular risk factors should be assessed and managed with awareness that typical anginal symptoms may not be present. Long-term complications for the ASO should be monitored for and patients should have routine follow-up at ACHD centers of excellence.

Compliance with Ethical Standards

Conflict of Interest
Lucy Safi declares no potential conflicts of interest.
Ami Bhatt is a section editor for *Current Treatment Options in Cardiovascular Medicine*.

Human and Animal Rights and Informed Consent
This article does not contain any studies with human or animal subjects performed by any of the authors.

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