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Arterial switch operation: A surgical triumph with long-term management challenges

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ABSTRACT

Since the late 1980s, the standard approach for treating D-transposition of the great arteries has been the arterial switch operation (ASO), replacing the Mustard/Senning procedure. Although ASO has shown impressive long-term survival rates, recent case series have revealed late complications such as neoaortic dilation and coronary artery stenosis. New findings emphasize the need for comprehensive evaluation of coronary risk and a deeper understanding of the mechanisms leading to coronary artery stenosis and myocardial ischemia over the long term. Computed tomography angiography (CTA) has unveiled a notable prevalence of abnormal coronary arteries with potential risk of stenosis and myocardial ischemia. Moreover, the progressive dilation of the neoaortic root and the potential for valve regurgitation necessitating intervention warrant serial imaging follow-up. Considering the radiation risks associated with CTA, magnetic resonance imaging emerges as a preferred modality for post-ASO patient assessment. Ongoing research in this field holds the promise of developing improved diagnostic and therapeutic strategies for these patients, thereby enhancing their long-term care

In 1975, Dr. Adib Dominos Jatene and colleagues pioneered a groundbreaking surgical procedure known as the arterial switch operation (ASO) to treat patients born with D-transposition of the great arteries (TGA) [1]. In the initial years following its introduction, the ASO was associated with a significant mortality rate. Nevertheless, during the 1980s, advancements in surgical techniques, perioperative medical care, coronary translocation, and the LeCompte manoeuvre ushered in a period of marked improvement in both short-term and long-term survival. These developments have underscored the ASO as a highly effective procedure.

The LeCompte manoeuvre, which involves the repositioning of the distal main pulmonary artery along with its branch pulmonary arteries to the forefront, while simultaneously shifting the distal aorta to a more posterior position, has emerged as the most employed technique for performing ASO [2]. In this procedure, the distal main pulmonary artery is anastomosed to the proximal aorta, connecting it with the right ventricle. Simultaneously, the distal ascending aorta is anastomosed to the proximal main pulmonary artery, creating a connection with the left ventricle outflow tract. Furthermore, the coronary artery ostia, originating from both the left and right sides, are meticulously excised from the native aortic root, along with the surrounding aortic walls. These carefully preserved components, often referred to as "buttons," are then translocated to the newly formed aortic root (neoaortic root).

Although the long-term outcomes of this surgical intervention are yet to be fully elucidated, numerous case series have reported remarkable long-term survival extending up to four decades post-ASO [3]. Within these case series, late complications have been noted, encompassing residual lesions such as neopulmonary and neoaortic valve regurgitation, supravalvar aortic and right ventricular outflow tract stenosis, progressive dilation of the neoaorta, and coronary artery stenosis.

Advancements in surgical techniques, notably the implementation of expansive trapdoor incisions to mitigate the extent of axial rotation of coronary buttons during translocation, have yielded tangible enhancements in surgical outcomes. This has, in turn, facilitated the effective transfer of all conceivable coronary patterns. Nevertheless, it is paramount to acknowledge that despite the feasibility of transitioning between different patterns, there undeniably exist certain native coronary dispositions that present more intricate technical challenges when it comes to the translocation process.

Consequently, the concern regarding an increased risk of coronary artery disease remains paramount, due to the associated risk of sudden cardiac death. In most TGA- ASO patients, one or both coronary arteries initiate their course between the great vessels. Their course may have a highly tangential orientation to the neo-aorta, with a plication of the proximal coronary's origin angle. However, there is no evidence for a correlation between the post-ASO coronary geometry and the likelihood of ischemia-related events.

In this issue of the Journal, Verheijen and colleagues have provided some elements for a screening strategy for coronary artery lesions in adults after ASO [4]. The authors are to be commended for their efforts. In the study, they included 81 adult TGA-ASO patients who underwent computed tomography angiography (CTA) and reported that 27% had an acute coronary take-off angle ($<30^\circ$), and 6% an inter-arterial course. Interestingly, patients who required coronary revascularization were among those with the most acute take-off angle ($<25^\circ$). This result underscores one of the possible mechanisms leading to coronary artery stenosis and myocardial ischemia among patients after ASO. Patients with acute coronary artery take-off angles may be more prone to late

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coronary compression due to overstretching of the coronary course as a result of progressive dilatation of the neoaortic root [5]. Another mechanism of coronary artery stenosis after ASO may be related to the positional changes of these arteries during growth. These changes may shift the transplanted coronary arteries (specially the main left and the left anterior descending coronary arteries) to a more anterior position, resulting in an interarterial course of these vessels and compression due to haemodynamic changes during exercise [6]. Furthermore, a prepulmonary course of coronary arteries in patients with a single coronary ostium may stretch the coronary arteries and lead to stenosis. An intramural course has been also described to be associated with a high risk of sudden cardiac death after ASO [7]. Finally, proximal eccentric intimal thickening has been observed and suggested as a potential mechanism of coronary stenosis after ASO [8].

The arrangement of great vessels and the positioning and the path of the transplanted coronary arteries post-ASO result in a condition akin to a surgically created anomalous aortic origin of the coronary arteries (AAOCA). Such as in AAOCA, several coronary lesions after ASO have led to ischemia-related ventricular arrhythmia [9]. Moreover, the surgical denervation of the coronary arteries may lead to both, a less laminar coronary blood flow, which may cause endothelial dysfunction and atherosclerosis over time, and a lesser vasodilatation response during exercise due to the lack of sympathetic activity [10].

Thus, we are left with a long-term management dilemma. What should we do with these patients? How should we follow them and how long? The 2018 AHA/ACC Guideline suggests periodic evaluation of coronary arteries using CTA, magnetic resonance imaging (MRI), or catheter angiography for asymptomatic patients to check coronary patency [11]. Further assessment is advised if symptoms of coronary ischemia arise. In asymptomatic individuals, the ESC guidelines question the indication for routine screening for coronary artery disease, whatever the modality [12]. The study of Verheijen et al. supports a systematic CTA screening of coronary arteries lesion at least once during the follow-up in adults after ASO, aiding in the early detection of high-risk coronary lesions, which will require additional evaluations, such as non-invasive myocardial ischemia testing. Because an increased incidence of cancer has been observed after diagnostic ionizing radiation exposure, including low-dose ionizing radiation exposure, magnetic resonance imaging for the study of the coronary arteries after ASO should be preferred over coronary computed tomography angiography [13]. However, detection of ischemia in such coronary artery anomalies is challenging as the mechanisms leading to ischemia differ from patients with coronary atherosclerosis. Stress testing modalities like MRI or positron emission tomography using physical exercise or dobutamine should probably be considered [14]. The study's findings also highlight a notably higher prevalence of cardiovascular risk factors in patients requiring reintervention after ASO, compared to those without such issues (40% vs. 7%). This result emphasizes the importance of implementing preventive measures for atherosclerotic cardiovascular disease to alleviate the acquired cardiovascular burden in these patients [15].

Another important finding from the Verheijen et al. study is the high prevalence of neoaortic dilatation (43%), including 10% who required neoaortic reintervention in a relatively young population (median age 21 years). They reported a progression of neoaortic root dilatation with age in adult TGA-ASO, with an average annual growth rate in neoaortic root dimensions of 0.16 mm over an 8-year follow-up period. Even though neoaortic root dilatation rarely progresses to neoaortic root dissection, baseline and serial imaging with echocardiography and MRI seems reasonable to confirm the absence of neoaortic dilatation. Five percent of patients from the study experienced neoaortic valve replacement [4]. The potential for neoaortic valve regurgitation seemed to increase as time passed post-surgery. As consequence, the initial severity of neoaortic insufficiency should determine the frequency of follow-up surveillance.

Even with the potential disease burden, most TGA-ASO patients have achieved remarkable long-term outcomes. The ongoing journey of post-ASO care can be viewed from different angles, much like the age-old question of whether the glass is half-empty or half-full. Therefore, by conducting high-quality research in the field (such as the current study) we may be able to design better diagnostic and therapeutic strategies, which may finally lead to improved patient outcomes and to tilt the scales in favour of a rather full glass.

Disclosure

One or more of the co-authors serve as members of the Editorial Board for IJC Congenital Heart Disease. They had no role in the editorial review of or decision to publish this article.

CRediT authorship contribution statement

Magalie Ladouceur: Conceptualization, Formal analysis, Writing – original draft. **Francisco Javier Ruperti-Repilado:** Conceptualization, Formal analysis, Writing – review & editing. **Tobias Rutz:** Formal analysis, Writing – review & editing.

Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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