

# The Arterial Switch Operation: A Tale of Great Expectations

*Cirugía de switch arterial: una historia de grandes esperanzas*

MARSHALL L. JACOBS<sup>1</sup>

Forty years ago, when Adib Jatene, in Sao Paulo, Brazil performed the first successful arterial switch operation on an infant with transposition of the great arteries and ventricular septal defect, it was celebrated as an outstanding technical achievement. Jatene had studied the published literature concerning transposition of the great arteries, and had become convinced that the problem of ventriculo-arterial discordance could be solved by “switching” the flow at the level of the great arteries if the coronary arteries could be successfully transposed, as had been suggested by Dr. Harold Albert. After reviewing many dozens of pathologic specimens of hearts with transposition, Jatene was struck by the consistent observation that the coronary arteries arose from the aortic sinuses adjacent to the pulmonary artery, and he was convinced that transfer of the coronary arteries with part of the surrounding sinus of Valsalva tissue would be possible and would require little mobilization. Though the first child on whom Jatene performed a “switch” procedure died at the end of the operation, he remained convinced that the principle was sound and that the operation was feasible. The second operation, on a 40-day-old child with transposition of the great arteries and a ventricular septal defect, was successful. (1) Cardiac surgery had witnessed a milestone achievement, and the field would change dramatically. Today, the Jatene operation is performed at virtually all pediatric cardiac surgery centers, and for many surgeons and centers the outcomes from arterial switch surgery serve as a yardstick by which to measure individual and team performance. With adoption of the arterial switch operation in the first few weeks of life for transposition with intact ventricular septum, and over a slightly wider range of age for transposition with hemodynamically important ventricular septal defect, as well as for the Taussig-Bing anomaly (actually a form of double outlet right ventricle), the arterial switch operation has become the mainstay of contemporary surgical management of several of the most common and most challenging forms of congenital heart disease.

The history of surgery for transposition was al-

ready a story of numerous fascinating innovations before the advent of anatomic repair. Varco and Lillehei, Baffes, and others had devised ingenious “closed heart” operations which achieved partial reversal of the systemic and pulmonary venous return to the atria. With refinement of the heart-lung bypass machine and application of cardiopulmonary bypass support to the youngest patients, the atrial baffle procedures developed by Senning and by Mustard were successfully performed by surgeons all over the world. By the time that arterial switch surgery had first been performed successfully, routine repair of simple and complex forms of transposition by means of atrial baffle repairs was commonplace, and the operations were being performed earlier and earlier in life, including routine repair of simple transposition in the newborn period at some centers.

Why then, make the “switch to the switch?” Data on short-term and intermediate-term survival after Mustard and Senning operations were generally favorable. Technical improvements led to reduction in the incidence of important obstructions in the systemic and pulmonary venous pathways. There was, however, a troubling incidence of late rhythm disturbances, and increasing concern as to whether the right ventricle and the tricuspid valve could function adequately in the systemic circulation over a normal lifespan. These concerns, and the evidence that supported them, seemed even more compelling with respect to the population of patients with transposition and ventricular septal defect. So, the arterial switch operation, and whatever “learning curve” and associated incremental risk of operative mortality might be experienced in the early phase after its adoption, began gradually to supplant other surgical strategies as the primary approach to repair of transposition of the great arteries and related anomalies. Multi-institutional studies, such as those of the Congenital Heart Surgeons’ Society in North America provided evidence to support this transition. (2, 3) The recognition that the natural “preparedness” of the left ventricle to assume the work of supporting the systemic circulation had a time-span that may be limited to the

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Address for reprints: Marshall L. Jacobs MD - Division of Cardiac Surgery - The Johns Hopkins Hospital - Zayed 7107, 1800 Orleans Street, Baltimore, MD 21287, USA - e-mail: [marshall.jacobs@comcast.net](mailto:marshall.jacobs@comcast.net)

<sup>1</sup> Division of Cardiac Surgery - Johns Hopkins University School of Medicine - Baltimore, Maryland, USA

first few weeks of life, led to the routine performance of arterial switch repair for transposition with intact ventricular septum in the neonatal period. Demonstration of the feasibility and short-term success of this strategy led also to earlier and earlier surgery for more complex forms of transposition.

There were, of course, many concerns that went beyond the challenge of the technical exercise itself. Would the “transplanted” coronary arteries function normally, and predictably? Would that be equally true regardless of the many different patterns of coronary origin and branching that are observed in patients with transposition? Would they be especially prone to coronary occlusive disease as a result of either surgical scarring or eventual atherosclerosis? Would the divided and “switched” great vessels exhibit appropriate growth at the level of the circumferential suture lines? What expectations were there with respect to the long-term durability of the native pulmonary valve in its new role as “neo-aortic” valve? And would this alternative to physiologic repair actually live up to predictions with respect to lower incidence of rhythm disturbances and durable functional status?

In this issue of *Revista Argentina de Cardiología*, Villalba and Capelli et al. present a review of their institutional experience with arterial switch surgery for transposition of the great arteries and a detailed analysis of follow-up data pertaining to 224 patients operated on with the arterial switch technique at Hospital de Pediatría “Prof. Dr. Juan P. Garrahan” from January 1992 to December 2013. (4) The cohort includes the full spectrum of variants (simple TGA, complex TGA with VSD and/or aortic arch obstruction, and the Taussig-Bing anomaly). Patients who died in the immediate postoperative period or were lost to follow-up (n=57) were excluded from the study. With respect to this being a study of intermediate and long-term status conditional upon initial operative survival, this is similar to some comprehensive single center studies, but differs from some others. In subtle ways, the exclusion of operative mortality may result in some mild, unintended bias in terms of the characteristics of the cohort. For example, the authors report that more than three-fourths of patients had the “normal” (or most common) coronary artery pattern. This is slightly more than the fraction of roughly two-thirds that is frequently observed in clinical or pathologic series. Whether this is related to a possible interaction between coronary anatomy and operative survival, or purely a matter of chance with respect to those who survived surgery but are lost to follow-up, or whether it is simply a reflection of the variation between one series and another is not knowable. It is unlikely however, that such subtle variations have important implications for the inferences drawn from the analysis.

With respect to the questions that have time and again been asked about long-term concerns regarding those who have survived arterial switch surgery early in infancy, this large experience from Buenos Aires joins a growing body of literature that is mostly reas-

suring. Mortality during follow-up (mean of 7.6 years) is just 1% (two patients) and all surviving patients are in Functional Class I-II. Importantly, all have normal ventricular function. As in other reports, the most common late complication is pulmonary stenosis (PS) at some level (17.4%), with supralvalvar stenosis being the most common site of obstruction. The observation that development of PS was associated with both the era of surgery and the duration of follow-up leaves one uncertain as to whether evolution of the surgical technique has relegated the highest likelihood of developing PS to those who underwent surgery in the earlier phase of the center’s experience, or alternatively, whether there is some degree of “inevitability” to this problem, and that those operated in the more recent epoch are still at risk for further late acquisition of PS. As in reports from other centers, neo-aortic root dilation (12% of patients) and time-related development of aortic valve regurgitation are clearly related to one another. Aortic regurgitation was associated with complex transposition and Taussig-Bing anomaly, prior left ventricular preparation, aortic root dilation, and presence of aortic regurgitation in the immediate postoperative period. Only five patients have undergone aortic valve replacements, including two composite root replacements (Bentall) and one “switch-back” Ross operation. Reoperations and catheter interventions directed at the right side of the heart were more numerous, as is true in most series. At 5, 10 and 15 years, 94%, 86% and 58% of patients, respectively, remained free from cardiac re-interventions of any type. Of five patients in whom there was evidence at any time of coronary artery problems, two presented within the first weeks or months following surgery, both with ventricular dysfunction. Three patients had late findings of coronary obstruction that was found during cardiac catheterization. But catheterization was not routinely performed as part of a follow-up protocol. One patient had additional residual lesions that were not related to the coronary issues, and did not survive a reoperation. The two remaining patients are asymptomatic, in FC I and with preserved ejection fraction.

With this analysis, Villalba et al. (4) have added to a rapidly growing body of information that one hopes will ultimately help us to answer questions about expectations regarding the long-term status of patients who have survived arterial switch repairs early in life. We know from this report and others that these patients are at risk for development of pulmonary outflow obstruction at various levels, and we suspect that improved surgical reconstructive strategies at the time of arterial switch surgery are likely to be associated with a reduction in the incidence of such problems, but it is doubtful that they will be eliminated entirely. It appears that aortic root dilation is related to identifiable risk factors including, among others, initial size discrepancy of the great vessels and pulmonary artery banding. Of patients who do develop aortic root dilation, further progression of dilation (in-

creasing Z-score over time) is observed in some, but certainly not in all. And the factors that influence progression are incompletely understood. Most late interventions directed at the neo-aortic root are indicated on the basis of aortic valve regurgitation, as opposed to isolated root dilation. Risk factors for time-related development of significant aortic valve regurgitation include older age at arterial switch surgery, TGA with VSD, initial aortic/pulmonary size discrepancy, aortic arch anomalies, and presence of aortic regurgitation at discharge following arterial switch operation. (5, 6) The incidence of arrhythmias in this and other arterial switch cohorts is low, but does include some patients with complex ventricular arrhythmias in the absence of an apparent anatomical substrate. Perhaps most concerning, is the fact that coronary obstructive lesions (2% in this series; 1-8 % in other reports) while uncommon, are often clinically silent, and frequently are associated with preserved ventricular function, at least for a period of time. One must ask whether there would be value in subjecting asymptomatic patients to periodic investigation of the status of the coronary arteries, and whether such surveillance would result in detection of obstructive lesions that have the potential to be the basis for future ischemic events. The significance of such obstructive lesions is incompletely understood. Many such lesions are likely to go undetected in the absence of a program of routine control cardiac catheterization or some alternative type of imaging and functional assessment of comparable yield and predictability.

Studies such as this one, which examine and analyze late postoperative events, are important in terms

of optimizing the management of enrolled patients. But they also help us to build an understanding of the “unnatural” history of patients following arterial switch surgery. That understanding is essential to our ability to formulate plans for rational practices in terms of routine surveillance and the timing of serial investigations. That’s an important part of enabling patients with transposition to achieve the maximal benefit from anatomic repair.

#### Conflicts of interest

None declared.

(See authors’ conflicts of interest forms in the website/Supplementary material).

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