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Left Ventricular Retraining: Theory and Practice

Richard G. Ohye, Ming-Sing Si, Edward L. Bove, and Jennifer C. Hirsch-Romano

Congenitally corrected transposition of the great arteries or l-transposition of the great arteries is characterized by discordance of both the atrioventricular and ventriculoarterial connections. Physiologic repair of associated conditions, whereby the morphologic right ventricle remains the systemic ventricle, has resulted in unsatisfactory long-term outcomes due to the development of right ventricular failure and tricuspid valve regurgitation. While intuitively attractive, anatomic repair also has inherent challenges and risks, particularly for those patients who present late and require left ventricular retraining. Although early and intermediate-term outcomes for anatomic repair have been encouraging, longer-term follow-up has demonstrated concern for late left ventricular dysfunction in this subgroup of patients. Continued monitoring of this challenging patient population will clarify late outcomes and inform clinical management in the future.

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Congenitally corrected transposition of the great arteries (CCTGA), or l-transposition of the great arteries, is an uncommon congenital cardiac malformation characterized by discordant atrioventricular and ventriculoarterial connections. It is commonly associated with additional cardiac defects, particularly ventricular septal defect (VSD) and pulmonary stenosis or atresia. Symptoms associated with CCTGA are generally related to one or a combination of these coexisting cardiovascular malformations. However, repair of the associated lesions, leaving the morphological right ventricle (RV) as the systemic ventricle (so called "physiologic repair"), as is also the case in isolated CCTGA, results in a decreased life expectancy caused primarily by the development of RV dysfunction and tricuspid valve regurgitation.[1] Reported survival for CCTGA has been 84% at 1 year, falling to 60% at 15 years.[1,2] True anatomic repair can be established by a double switch technique, which combines an atrial-level switch (Senning or Mustard) with an arterial-level switch (arterial switch operation, Rastelli or Nikaidoh). Many authors reserve the term "double switch" specifically for the combination of a Senning or Mustard and an arterial switch operation. For the purposes of this review, the term "double switch" will encompass any combination of atrial and arterial-level switches. Several centers have reported favorable short- and intermediate-term results of patients undergoing anatomic repair of CCTGA.[3–8]

Unless there is a non-restrictive VSD and/or pulmonary outflow tract obstruction, the left ventricle (LV) will become deconditioned in the first few weeks following birth in patients with CCTGA. As with d-transposition of the great arteries (d-TGA), performing a double switch operation in a patient with CCTGA after the LV is deconditioned will result in LV failure when the ventricle is required to acutely generate systemic pressures. For CCTGA patients with moderate pulmonary outflow tract obstruction, or those with a VSD and pulmonary outflow tract obstruction who are well balanced, the LV remains conditioned by the systemic or near-systemic pressures. These patients may be simply observed closely as an outpatient for the development of complications, such as RV dysfunction or tricuspid valve regurgitation. For other patients, there will be a VSD without the proper degree of pulmonary outflow tract obstruction, resulting in congestive heart failure and the need for a pulmonary artery band (PAB). For newborns with CCTGA without associated defects, but who demonstrate evidence of dysfunction and tricuspid valve regurgitation, it has been our approach to place a relatively loose PAB within the first few weeks of life. This approach was proposed by Yacoub et al[9] in 1977 as a means to delay the need to perform the arterial switch operation for d-TGA beyond the neonatal period. Those patients with pulmonary atresia or severe pulmonary outflow tract obstruction will require systemic-to-pulmonary artery shunting. We have generally

Section of Pediatric Cardiovascular Surgery, Department of Cardiac Surgery, University of Michigan Medical School, Ann Arbor, MI.

Dr. Ohye reports receiving consulting fees from Sorin and CryoLife.

Address correspondence to: Richard G. Ohye, MD, Room 11-742, 1540 E. Hospital Drive/SPC 4204, Ann Arbor, MI 48109-4204. E-mail: ohye@umich.edu

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KEY POINTS

- Unsatisfactory long-term results from physiologic repair of CCTGA has led to an interest in anatomic repair.
- A cohort of patients with CCTGA will require LV retraining prior to attempted anatomic repair.
- LV retraining for CCTGA is possible in many cases, but may be of more limited success in the patients beyond late childhood.
- Although early and intermediate-term outcomes for anatomic repair have been encouraging, longer-term follow-up has demonstrated concern for late left ventricular dysfunction in patients requiring LV retraining.

preferred to wait until at least 6 months of age before undertaking the double switch, although our youngest patient was 6 weeks of age at repair.[4]

For those patients presenting late with a deconditioned LV, the LV must be retrained in preparation for a double switch. This reconditioning is done by increasing the afterload on the LV with a PAB. This concept was initially described by Mee[10] in 1986 as a means to retrain the morphologic LV in patients with d-TGA to allow a late arterial switch operation for patients who had previously undergone an atrial-level switch. For neonates, our institutional approach is to place a relatively loose band, perhaps at 50% systemic systolic pressure, to allow for growth. For older patients, the desired gradient across the PAB depends primarily on the amount of pressure increase the LV will acutely tolerate without demonstrating dysfunction. The gradient and the function of the LV are assessed using transesophageal echo and direct pressure measurements. Some patients may be clinically improved by the placement of the PAB alone because the increased LV pressure results in a rightward shift of the ventricular septum, decreasing the degree of tricuspid regurgitation. In some patients who do not tolerate a tight band, it may be necessary to progressively tighten the band over time to eventually achieve a goal of 75% to 80% of systemic pressure before embarking on a double switch. Various techniques of adjustable PABs have been described that facilitate adjustments of the band over time without the need for resternotomy.[11–19] Once the pressure in the morphologic LV is 75% to 80% of systemic associated with normal LV function, it is generally believed to be prepared for the systemic workload. Other indicators of LV preparedness include normalized LV mass and wall thickness by MRI or echocardiography. Some centers will routinely perform cardiac catheterization for follow-up, others will perform a catheterization to confirm adequate LV pressures before undertaking the double switch, and still others will challenge the patient with isoproterenol to determine if the LV can generate suprasystemic pressures as a prerequisite to proceeding with the double switch. While there are anecdotal reports of adults undergoing successful LV reconditioning followed by a successful double switch operation,[20] the literature would suggest that patients in their adolescence and later are highly unlikely to tolerate satisfactory LV retraining. This is due, at least in part, to the fact that immature hearts will respond to a PAB with both cellular hyperplasia and hypertrophy, whereas older hearts are limited to the development of hypertrophy only and are, therefore, susceptible to develop diastolic dysfunction as the ventricular wall hypertrophies.

The early and intermediate-term results of the double switch operation have been favorable. Our institution reported an experience with 35 consecutive patients undergoing a double switch over approximately a 10-year period.[4] Twenty-three subjects underwent double switch because of RV dysfunction with tricuspid valve regurgitation (n=15) or associated uncorrected cardiac lesions (n=8). PAB was necessary in 15 subjects for LV retraining (n=11) or for congestive heart failure (n=4). Of the 15 subjects requiring LV retraining, two patients, aged 12 and 14 years, failed retraining because of LV dysfunction. One subject, who underwent LV reconditioning and subsequent double switch at 7 years of age, required cardiac transplantation for progressive LV failure. Four subjects remained banded, awaiting repair. At the time of that report, 17 subjects had undergone a double switch with no early or late mortality at a mean follow-up of 36 months (range, 1 month to 8 years).

There have been more recent concerns surrounding the long-term functioning of the retrained LV. Animal models have demonstrated that retrained ventricles display subendocardial edema, myocardial necrosis, and fibrosis with reduced ventricular work index.[21,22] Brawn and colleagues[23] published their late results in 44 subjects, comparing those who required LV retraining (n=11) and those who did not (n=33). The 30-day hospital mortality was two of 44 subjects (4.5%) and the long-term raw mortality was five of 44 (11%). Rate of death and transplantation combined was seven of 44 (16%). The actuarial freedom from death, transplantation, or the development of moderate-to-severe LV dysfunction was 85% at 1 year, 80% at 5 years, and 72% a 10 years. Comparing groups, there were no significant differences in mortality or the incidence of death/transplantation. Yet, while the differences did not reach statistical significance, the incidence of moderate-to-severe LV dysfunction at 1 year following double switch was 39% in subjects requiring LV retraining, compared with 6% of subjects not requiring retraining. In addition, when death, transplantation, and moderate-to-severe LV dysfunction were analyzed as a composite endpoint, those subjects undergoing LV retraining before double switch had a statistically worse outcome than those not requiring retraining (P =.04).

While most experienced centers have adopted an approach of anatomic repair for CCTGA, more data are needed to define the role for LV retraining in older patients, as well as to assess late outcomes in all patients undergoing a double switch. Further long-term follow-up will better define which patients benefit most from anatomic repair, physiologic repair, or cardiac transplantation.

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