Late left ventricular dysfunction after anatomic repair of congenitally corrected transposition of the great arteries

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Objective: Early results for anatomic repair of congenitally corrected transposition of the great arteries (ccTGA) are excellent. However, the development of left ventricular dysfunction late after repair remains a concern. In this study we sought to determine factors leading to late left ventricular dysfunction and the impact of cardiac resynchronization as a primary and secondary (upgrade) mode of pacing.

Methods: From 1992 to 2012, 106 patients (median age at surgery, 1.2 years; range, 2 months to 43 years) with ccTGA had anatomic repair. A retrospective review of preoperative variables, surgical procedures, and postoperative outcomes was performed.

Results: In-hospital deaths occurred in 5.7% (n = 6), and there were 3 postdischarge deaths during a mean follow-up period of 5.2 years (range, 7 days to 18.2 years). Twelve patients (12%) developed moderate or severe left ventricular dysfunction. Thirty-eight patients (38%) were being paced at latest follow-up evaluation. Seventeen patients had resynchronization therapy, 9 as an upgrade from a prior dual-chamber system (8.5%) and 8 as a primary pacemaker (7.5%). Factors associated with left ventricular dysfunction were age at repair older than 10 years, weight greater than 20 kg, pacemaker implantation, and severe neo-aortic regurgitation. Eight of 9 patients undergoing secondary cardiac resynchronization therapy (upgrade) improved left ventricular function. None of the 8 patients undergoing primary resynchronization developed left ventricular dysfunction.

Conclusions: Late left ventricular dysfunction after anatomic repair of ccTGA is not uncommon, occurring most often in older patients and in those requiring pacing. Early anatomic repair and cardiac resynchronization therapy in patients requiring a pacemaker could preclude the development of left ventricular dysfunction. (J Thorac Cardiovasc Surg 2014;148:254-8)

CHD

Congenitally corrected transposition of the great arteries (ccTGA) is a complex cardiac anomaly representing approximately 1% of all congenital heart defects.[1] Classic or physiologic repair of this condition has been possible since the late 1950s.[2] However, in patients undergoing a physiologic approach, as well as in patients not undergoing surgery, the morphologic right ventricle (RV) remains connected to the systemic circulation, leading to late RV dysfunction and tricuspid regurgitation with disappointing late results. In fact, systemic ventricular failure may affect as many as 40% of patients within 3 years of physiologic repair, and 56% of unoperated patients older than age 45 years.[3,4]

The guarded outcomes of the morphologic RV in the systemic circulation provided the impetus for the surgical management of ccTGA to evolve from physiologic to anatomic repair, which restores the morphologic left ventricle (LV) into the systemic circulation.[5,6] The so-called double switch entails an atrial switch (Senning or Mustard procedure), and an arterial switch, Rastelli procedure, or, more recently, an aortic translocation procedure to restore the atrioventricular and ventricle-arterial concordance.[7,8] Excellent results of this approach have been reported consistently since the mid-1990s and, therefore, this approach has been widely adopted.[9] Nevertheless, the development of late morphologic LV dysfunction still develops in a significant proportion of patients undergoing anatomic repair.[10]

Neo-aortic regurgitation (neo-AR) and previous left ventricular (LV) retraining with pulmonary artery banding (PAB) have been reported as determinants of LV dysfunction after anatomic repair of ccTGA.[11,12] We have previously published a higher incidence of left ventricular dysfunction in those patients requiring pacing and with prolonged QRS for age and suggested that cardiac resynchronization therapy (CRT) could be of value in this population.[10] However, other factors contributing to LV dysfunction are poorly understood. Management of LV dysfunction in patients undergoing anatomic repair of ccTGA is controversial.

We report our experience with anatomic repair of ccTGA in 106 consecutive patients, especially focusing on late outcomes, in particular the development of late LV

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication June 18, 2013; revisions received Aug 8, 2013; accepted for publication Aug 16, 2013; available ahead of print Oct 7, 2013.

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0022-5223/$36.00

http://dx.doi.org/10.1016/j.jtcvs.2013.08.047

Abbreviations and Acronyms

ccTGA = congenitally corrected transposition of the great arteries
CRT = cardiac resynchronization therapy
LV = left ventricle
neo-AR = neo-aortic regurgitation
PAB = pulmonary artery banding
RV = right ventricle

dysfunction. Factors associated with this complication and management strategies are presented.

MATERIALS AND METHODS

Study Population

From June 1992 through June 2012, 106 consecutive patients, 67 males (63.2%), with ccTGA underwent an anatomic repair. The median age at surgery was 1.2 years (range, 2 months to 43 years), and the median weight was 9.4 kg (range, 3.4-58.5 kg). The segmental anatomy was S, L, L in 93 patients (87.7%) and I, D, D in 13 patients (12.3%). Levocardia was present in 69 patients (65.0%), dextrocardia was present in 26 patients (24.5%), and mesocardia was present in 11 patients (10.5%). Associated anomalies included ventricular septal defects in 93 patients (87.7%), pulmonary stenosis in 42 patients (39.6%), pulmonary atresia in 31 patients (29.2%), and Ebsteinoid dysplasia of the left-sided tricuspid valve in 23 patients (21.7%).

Surgical Technique

Before anatomic repair, 29 patients (27.4%) had a modified Blalock-Taussing shunt and 30 patients (28.3%) had PAB to retrain the LV or prevent overcirculation. An atrial switch was accomplished by a modified Senning procedure in 35 patients (33%) and a Mustard surgery in 71 patients (67%). Forty-two patients (39.6%) had a Rastelli procedure for LV outflow obstruction, and 62 (58.5) underwent an arterial switch for restoration of the ventriculo-arterial concordance. Two patients with severe subpulmonary stenosis (1.9%) had an aortic translocation procedure late in our experience. We have previously described in detail the procedures performed to restore the atrioventricular concordance (Mustard or Senning) as well as the ventriculoarterial concordance (arterial switch, and Rastelli and Nikaidoh procedures).[7,8] The surgical technique characteristics are shown in Table 1. Our protocol for CRT in pediatric patients also has been reported.[13]

Data Collection and Statistical Analysis

This study was performed at Children's Hospital in Boston and was approved by the Institutional Review Board. Data were obtained from a retrospective review of hospital charts and medical records preoperatively and postoperatively. Individual patient consent for this study was waived. The degree of LV dysfunction was determined by echocardiographic qualitative assessment before repair and serial follow-up echocardiograms. To confirm that LV dysfunction was present, magnetic resonance imaging was reviewed when available and/or the echocardiographic end-systolic wall stress to fractional-shortening relationship. In patients with LV dysfunction, coronary angiograms were reviewed when available. LV dysfunction was considered significant if graded as being moderate or severe. The degree of neo-AR was determined with Doppler echocardiography by the vena contracta jet width. In patients requiring a pacemaker and/or CRT implantation, electrocardiograms were reviewed.

Relationships between patients and surgical characteristics and the presence of significant LV dysfunction were examined using the Fisher exact test or the χ^2 test when appropriate; continuous variables, such as age at surgery, were categorized before the analyses were performed. A P value of .05 or less was considered significant.

RESULTS

In-hospital mortality rate was 5.7% (n = 6). One patient with preoperative severe tricuspid regurgitation went on extracorporeal membrane oxygenation after anatomic repair and successfully was bridged to heart transplantation. The late mortality rate was 3% (n = 3). One patient died of leukemia, another patient with pulmonary atresia and multiple aortic to pulmonary artery collaterals died of right ventricular failure, and the last patient had severe neo-aortic regurgitation and LV dysfunction. Freedom from death or transplantation is shown in Figure 1.

Excluding the 6 early deaths and for a mean follow-up period of 5.2 years (range, 7 days to 18.2 years), 12 patients (12%) developed significant LV dysfunction. A complete or high-degree heart block requiring a pacemaker implantation developed in 15 patients preoperatively, in 12 patients at surgery, and in 11 patients postoperatively. Of the total 38 patients requiring a pacemaker, 12 patients developed postoperative LV dysfunction during follow-up evaluation. CRT was performed in 17 patients, 9 as an upgrade from a previous dual-chamber system and 8 as a primary pacing system. LV function on late follow-up evaluation was normal in 70 patients, mildly depressed in 18 patients, and significantly (moderately or severely) depressed in 12 patients. With respect to CRT, LV function improved in 8 of 9 patients undergoing a CRT upgrade. Moreover, none of the patients undergoing CRT as their primary pacing system developed LV dysfunction through the follow-up period. Of the 30 patients undergoing PAB before anatomic repair, 4 patients developed LV dysfunction during follow-up evaluation. Two patients in the arterial-switch group developed significantly neo-AR and LV dysfunction. One of those patients with a pacemaker died suddenly at home, and the other patient underwent a neo-aortic valve replacement. Interestingly, neither of the patients had undergone previous PAB. In univariate analysis, late LV dysfunction was related significantly to weight greater than 20 kg or age older than 10 years at anatomic repair, severe neo-aortic valve regurgitation, and a ventricular pacemaker. PAB did not come up as a significant factor for this late complication. No risk factor for LV dysfunction was found in multivariate analysis. An analysis of potential risk factors for late LV dysfunction is shown in Table 2.

DISCUSSION

Anatomic repair of ccTGA provides an excellent alternative to traditional or physiologic surgical management. The reported low surgical morbidity and mortality of anatomic repair of ccTGA, combined with its good midterm outcomes, have extended the applicability of the procedure to