The hemi-Mustard/bidirectional Glenn atrial switch procedure in the double-switch operation for congenitally corrected transposition of the great arteries: Rationale and midterm results

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Objective: This study was undertaken to assess the risks and benefits of the double-switch operation using a hemi-Mustard atrial switch procedure and the bidirectional Glenn operation for congenitally corrected transposition of the great arteries. To avoid complications associated with the complete Senning and Mustard procedures and to assist right-heart hemodynamics, we favor a modified atrial switch procedure, consisting of a hemi-Mustard procedure to baffle inferior vena caval return to the tricuspid valve in conjunction with a bidirectional Glenn operation.

Methods: Between January 1994 and September 2009, anatomic repair was achieved in 48 patients. The Rastelli–atrial switch procedure was performed in 25 patients with pulmonary atresia and the arterial–atrial switch procedure was performed in 23 patients. A hemi-Mustard procedure was the atrial switch procedure for 70% (33/48) of anatomic repairs.

Results: There was 1 in-hospital death after anatomic repair. There were no late deaths or transplantation. At a median follow-up of 59.2 months, 43 of 47 survivors are in New York Heart Association class I. Bidirectional Glenn operation complications were uncommon (2/33), limited to the perioperative period, and seen in patients less than 4 months of age. Atrial baffle–related reoperations or sinus node dysfunction have not been observed. Tricuspid regurgitation decreased from a mean grade of 2.3 to 1.2 after repair ($P = .00002$). Right ventricle–pulmonary artery conduit longevity is significantly improved.

Conclusions: We describe a 15-year experience with the double-switch operation using a modified atrial switch procedure with favorable midterm results. The risks of the hemi-mustard and bidirectional Glenn operation are minimal and are limited to a well-defined patient subset. The benefits include prolonged conduit life, reduced baffle- and sinus node–related complications, and technical simplicity. (J Thorac Cardiovasc Surg 2011;141:162-70)

Surgical management of congenitally corrected transposition of the great arteries (ccTGA) has evolved from a physiologic repair that addresses structural lesions to an anatomic approach that supports the systemic circulation with the morphologic left ventricle (LV). Adoption of the double-switch operation has been driven by the dismal natural history of the systemic right ventricle (RV). Progressive functional impairment of the systemic RV has been observed in 40% at 3 years and 60% at 10 years after traditional repair.[1,2] Successful early outcomes with anatomic repair were first reported by several centers in the mid-1990s.[3-5]

The anatomic variability observed in corrected transposition poses numerous technical challenges for anatomic repair. The surgical strategy must be tailored to account for associated cardiac lesions that are common in patients with ccTGA, such as ventricular septal defect (VSD), subpulmonary ventricular outflow tract obstruction or pulmonary atresia, and tricuspid valve dysplasia leading to incompetence of the systemic atrioventricular valve. Morphologic variables, including dextrocardia and atrial situs inversus, can greatly affect the visualization to create the atrial baffle.

It has been our preference to use a hemi-Mustard procedure that baffles the inferior caval return to the tricuspid valve in conjunction with a bidirectional Glenn (BDG) operation for the atrial switch component of the double-switch operation. Historical experience with the Mustard and Senning atrial switch procedures has identified a significant incidence of systemic venous and, to a lesser extent, pulmonary venous obstruction over the long-term.[6-8] Moreover, late development of sinus node dysfunction after the atrial switch procedure can be as high as 40% and is a risk factor for sudden death.[7,9,10] The hemi-Mustard

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Abbreviations and Acronyms

AAS = arterial–atrial switch
BDG = bidirectional Glenn
ccTGA = congenitally corrected transposition of the great arteries
NYHA = New York Heart Association
PA = pulmonary artery
PAB = pulmonary artery banding
RAS = Rastelli–atrial switch
RV = right ventricle
TR = tricuspid regurgitation
VSD = ventricular septal defect

modification obviates the need for the superior caval suture lines and, as a result, might result in a reduced incidence of these late complications of traditional atrial baffle procedures. Use of the BDG operation also decreases the volume load on the failing RV and reduces strain on a dysplastic or Ebsteinoid tricuspid valve, providing potential benefits. The reduced volume load should be particularly beneficial in those patients receiving an RV–pulmonary artery (PA) conduit.

An added benefit of the hemi-Mustard procedure is the technical advantage afforded in cases in which adequate exposure for construction of the atrial baffle is limited, such as dextrocardia or situs inversus. The posterior displacement of the atrial chambers in the setting of these positional anomalies combined with the lack of volume loading of the atria due to the absence of an atrial septal defect compromise visualization to perform the conventional Mustard or Senning baffle procedure.

This study represents a 15-year experience with anatomic repair for all cases of ccTGA with 2 adequate ventricles. When possible, the hemi-Mustard/BDG modified atrial switch procedure was performed along with an arterial switch or Rastelli procedure. This study focuses particularly on assessing the risks and benefits of this particular technique. Patient selection, operative strategy, and midterm outcomes are presented.

MATERIALS AND METHODS

Between January 1994 and September 2009, 48 patients with ccTGA and 2 adequately sized ventricles underwent a surgical management strategy designed to place the morphologic LV in the systemic circulation (Figure 1). A retrospective review of patients' charts, the operative record, diagnostic reports, and outpatient clinic records was performed in accordance with an accepted protocol from the Stanford University Institutional Review Board. Individual patient consent was waived because of the retrospective nature of the study.

Diagnosis of ccTGA and delineation of cardiac anatomy was established by means of echocardiographic analysis in all cases. Echocardiographic analysis also was used to characterize systolic ventricular function and the severity of tricuspid regurgitation (TR). TR was classified on a scale of 1 to 4 (1, trace; 2, mild; 3, moderate; and 4, severe).

Patient follow-up was obtained from records of the most recent pediatric cardiologist's examination and assessment. The most recent echocardiogram was obtained for assessment of ventricular function, valve function, and baffle leak or obstruction. Follow-up ranged from 7 months to 16 years, with a median follow-up of 4.9 years.

Patients' Characteristics

There was an equal distribution of male and female patients in this study (24 of each sex). The median age at anatomic repair was 3.0 years (range, 3.9 months–24.0 years). Ten patients were less than 1 year of age at time of the double-switch operation. The age distribution is depicted in Figure 2.

The most common associated cardiac defect was a VSD, which was present in 40 patients. Pulmonary atresia was present in 22 patients. Four of these patients had major aortopulmonary collateral arteries as the source of pulmonary blood flow. Severe subpulmonary obstruction resulting in functional pulmonary atresia was present in 3 patients. Seven patients had mild-to-moderate subpulmonary obstruction. Tricuspid valve abnormalities were present in 22 patients. In 10 patients the tricuspid valve had Ebsteinoid features. TR was at least moderate in 20 patients. Complete heart block requiring permanent pacing was present in 4 patients before anatomic repair.

Positional anomalies of the cardiac apex were common, occurring in 35% of patients in this series. Of the 48 patients undergoing the double-switch operation, dextrocardia with situs solitus was present in 10 (20%), and dextrocardia with situs inversus was present in 6 (13%). An additional patient had mesocardia with situs solitus.

Surgical Strategy

Anatomic correction for ccTGA was tailored to the patients' specific anatomies. Patients underwent one of 2 approaches to anatomic repair (1) an arterial–atrial switch (AAS) procedure if an appropriately sized pulmonary valve was present (n = 23) or (2) the Rastelli–atrial switch (RAS) procedure for patients with pulmonary atresia (n = 22) or severe subpulmonary stenosis (n = 3).

Surgical procedures performed before anatomic repair are displayed in Table 1. In the AAS group the majority of these procedures were designed to prepare the morphologic LV for the systemic afterload. Pulmonary artery banding (PAB) was performed in 17 (74%) patients before the AAS procedure. Eight of these patients required PAB tightening before anatomic repair because of inadequate LV preparation. The suitability of the AAS procedure after PAB was determined based on echocardiographic evidence of normal LV and mitral valve function, cardiac catheterization evidence of near-systemic morphologic LV pressure and low end-diastolic pressure, and achievement of normal LV mass by means of magnetic resonance imaging.

Patients undergoing the RAS procedure because of pulmonary atresia typically required a systemic artery–PA shunt to provide a reliable source of pulmonary blood flow. A modified Blalock–Taussig shunt was performed in 17 (68%) patients undergoing the RAS procedure. Of the 4 patients with pulmonary atresia and major aortopulmonary collateral arteries, 2 required an aortopulmonary window to promote growth of diffusely hypoplastic branch pulmonary arteries. All 4 patients ultimately underwent 1-stage unifocalization to a central aortopulmonary shunt before anatomic repair of ccTGA.

Surgical Technique

PAB. A midline sternotomy was performed to expose the aorta and PA. Minimal dissection was performed to encircle the proximal main PA with a Silastic band (Dow Corning, Midland, Mich). The degree of band tightening was guided by simultaneous transesophageal echocardiographic analysis and direct proximal PA pressure measurement. As the band was