Mid-Term Outcomes in Patients with Congenitally Corrected Transposition of the Great Arteries: A Single Center Experience

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BACKGROUND: Optimal management of patients with congenitally corrected transposition of the great arteries (ccTGA) is unclear. The goal of this study was to compare the outcomes in patients with ccTGA undergoing different management strategies.

STUDY DESIGN: Patients with ccTGA believed suitable for biventricular circulation, treated between 1995 and 2016, were included. The cohort was divided into 4 groups: systemic right ventricle (RV) (patients without surgical intervention or with a classic repair), anatomic repair, Fontan palliation, and patients receiving only a pulmonary artery band (PAB) or a shunt. Transplant-free survival from presentation was calculated for each group.

RESULTS: The cohort included 97 patients: 45 (46%) systemic RV, 26 (27%) anatomic repair, 9 (9%) Fontan, and 17 (18%) PAB/shunt. Median age at presentation was 2 months (range 0 days to 69 years) and median follow-up was 10 years (1 month to 28 years). At initial presentation, 10 (11%) patients had any RV dysfunction (8 mild, 2 severe), and 16 (18%) patients had moderate or severe tricuspid regurgitation (TR). During the study, 10 (10%) patients died, and 3 (3%) patients underwent transplantation. At last follow-up, 11 (11%) patients were in New York Heart Association class III/IV, 5 (5%) had moderate or severe systemic ventricle dysfunction, and 16 (16%) had moderate or severe systemic atrioventricular valve regurgitation. Transplant-free survivals at 10 years were 93%, 86%, 100%, and 79% for systemic RV, anatomic repair, Fontan palliation, and PAB/shunt, respectively ($p = 0.33$). On multivariate analysis, only systemic RV dysfunction was associated with a higher risk for death or transplant ($p = 0.001$).

CONCLUSIONS: Transplant-free survival in ccTGA appears to be similar between patients with a systemic RV, anatomic repair, and Fontan procedure. Systemic RV dysfunction is a risk factor for death and transplant. (J Am Coll Surg 2017;224:707–715. © 2017 by the American College of Surgeons. Published by Elsevier Inc. All rights reserved.)

Congenitally corrected transposition of the great arteries (ccTGA) is a complex cardiac anomaly representing 1% or less of all congenital heart defects.[1] The principal anatomic lesion in ccTGA is the presence of atrioventricular and ventriculoarterial discordance. The clinical presentation of patients with ccTGA is highly variable, ranging from asymptomatic patients to those with overt heart failure. This spectrum of clinical presentation is influenced by associated anomalies that may accompany ccTGA, such as ventricular septal defect (VSD), pulmonary stenosis or atresia, anatomic or functional abnormalities of the tricuspid valve, or anomalies in the conduction system, which have some effect on either the pulmonary circulation, or ultimately, influence ventricular function.[2]

Outcomes for these complex patients have not been well described, and the optimal management of patients

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Abbreviations and Acronyms

AVB = atrioventricular block
ccTGA = congenitally corrected transposition of the great arteries
NYHA = New York Heart Association
PAB = pulmonary artery band
RV = right ventricle
VSD = ventricular septal defect

with ccTGA still remains unclear.[3] Current strategies include a "classic repair," which addresses only the associated defects, leaving the morphologic right ventricle (mRV) in the systemic circulation, and has been found to be associated with progressive tricuspid regurgitation and dysfunction of the morphologic right ventricle in the long-term[4]; anatomic repair, which corrects both the atrioventricular and ventriculoarterial discordances as well as any associated defect; and single-ventricle palliation in the form of a total cavopulmonary connection for a subgroup of patients who may present with discordant connections and whose anatomy may not be suitable for biventricular repair.

Pulmonary artery banding (PAB) and systemic to pulmonary artery shunting have also been used as intermediate procedures in patients with ccTGA. The purpose of these strategies is to retrain the morphologic left ventricle (mLV) before an anatomic repair, protect the pulmonary vascular bed before a definitive surgical repair, and/or bridge the patient to a later point in life, when anatomic repair is conceivably less challenging from a technical standpoint. Further, there are patients whose clinical presentation is relatively uncomplicated, who may be managed medically without the need for surgical intervention and who remain with adequate right ventricular function well into adult life.[1]

The goal of this study was to compare the outcomes of patients with ccTGA undergoing different management strategies and identify potential risk factors for death or transplantation in a large volume single institution.

METHODS

The study cohort included all patients diagnosed with ccTGA, potentially suitable for biventricular circulation, who were treated at Texas Children's Hospital from 1995 to 2015. Suitability for biventricular repair was determined by reviewing the clinical consultation and multidisciplinary management conference notes. As such, patients with a plan that was clearly stated as going down a single-ventricle pathway were not included. The ultimate decision as to which type of repair to perform for these patients was made by the surgeon after reviewing all the data and/or performing intracardiac exploration or hemodynamic measurement at the time of surgery. Patients who had their definitive repair at an outside hospital and were treated at our institution afterward, were also excluded from the study. All demographic and clinical data were collected by retrospective review of all medical records, operative reports, procedure notes, discharge notes, and clinic notes. Follow-up was obtained by a combination of patient records and structured telephone interviews with patients and referring cardiologists or primary care physicians. This study was approved by the IRB at Baylor College of Medicine, and informed consent was waived.

The cohort was divided into 4 groups: systemic right ventricle (RV) (patients who were only followed medically, or those with a classic repair), anatomic repair (in the form of an atrial and arterial switch, or atrial switch-Rastelli), Fontan palliation (those patients who, at the time of repair, were found not to be candidates for biventricular circulation after intracardiac exploration because of a complex anatomy [remote VSDs, morphology of atrioventricular valve] that would complicate an anatomic repair), and patients receiving only a pulmonary artery band (PAB) or a shunt and were waiting for definitive repair. Patients who underwent pacemaker placement, but received no other surgical intervention, were included in the medical group category. Initial time was the patient's first encounter at our institution. Reintervention was defined as the need for catheter-based intervention (balloon angioplasty or stent placement), or surgical intervention after the definitive repair for patients in the classic, anatomic repair, and Fontan groups. Perioperative mortality was defined as death before hospital discharge or within 30 days of surgery.

Echocardiographic evaluations at first encounter and at last follow-up were reviewed to assess ventricular and atrioventricular valve function. Systemic ventricular function was defined qualitatively as normal or as mildly, moderately, or severely depressed. Systemic atrioventricular valve regurgitation was defined qualitatively as absent, mild, moderate, or severe. At last follow-up, in the anatomic repair group, ventricular function and atrioventricular valve regurgitation were defined by the left ventricle and mitral valve, respectively.

Data analysis

Descriptive analyses were performed for the entire cohort and for each of the groups. Data are described as percentages and medians with ranges, as appropriate. Univariate analyses for transplant-free survival and freedom-from-reintervention were performed using the Kaplan-Meier

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