Heart Transplantation in Adults With Congenital Heart Disease: 100% Survival With Operations Performed by a Surgeon Specializing in Congenital Heart Disease in an Adult Hospital

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Background. Cardiac transplantation in adult patients with congenital heart disease poses numerous challenges. The optimal operative and postoperative management strategies remain unclear. The purpose of our study was to (1) characterize the adult patient with a congenital heart condition undergoing transplantation, the operation, and the postoperative course; (2) report the survival after heart transplantation at our center; and (3) discuss issues surrounding the unique setting we provide for the operative and postoperative care of this complex patient cohort.

Methods. We performed a retrospective cohort study of 12 consecutive adult patients with a prior history of congenital heart disease who underwent heart transplantation at a single, large, academic center between September 1, 2005, and September 1, 2013. The operations were performed by a surgeon specializing in congenital heart disease in an adult hospital. Postoperative care was provided jointly by that surgeon and the adult cardiac transplantation team.

Results. At operation, the median age and weight were 41 years (range, 16 to 72 years) and 65 kg (range, 45 to 104 kg), and 100% of patients had undergone previous operations. The median donor ischemic time was 197 minutes (range, 137 to 282 minutes). The median cardiopulmonary bypass time was 210 minutes (range, 175 to 457 minutes), and the median total operating time was 582 minutes (range, 389 to 968 minutes). Three patients required mechanical support to be weaned from cardiopulmonary bypass. Postoperatively, 3 patients required the addition of mechanical support in the intensive care unit, and 3 patients required tracheostomy for prolonged ventilation. The majority of patients had a complicated postoperative course (66%). The median number of noncardiac consultants required to help care for these patients was four (range, two to 12). The mortality was 0%.

Conclusions. Cardiac transplantation in adults with congenital cardiac disease is challenging, is fraught with adverse events, and requires meticulous care and teamwork for success. A surgeon specializing in congenital heart conditions may be best to handle the operative challenges, and an adult hospital with access to certain technology and consultant services may be best to handle the postoperative challenges in this difficult patient population.

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Patients with congenital heart disease experience inferior short-term survival after orthotopic heart transplantation compared with patients undergoing transplantation for noncongenital pathologic conditions. The data show a 1-month survival of 86%, the lowest rate among the analyzed subpopulations (congenital, cardiomyopathy, valvular, coronary artery disease, and retransplantation) [1].

These transplant operations are often technically challenging because of anatomic constraints and prior congenital heart operations. The postoperative courses can also be complicated, requiring consultations from numerous subspecialists. Our operations in adults with congenital heart disease, including heart transplantations in these adults, are performed by a surgeon specializing in congenital heart conditions within an adult hospital. We have demonstrated that this system may offer the patients the greatest opportunity for survival [2].

The purpose of our study was to (1) characterize the adult patient with congenital heart disease undergoing transplantation, the operation, and the postoperative course; (2) report the survival after heart transplantation at our center; and (3) compare our operative and postoperative care environment with that of other centers in the nation.

Patients and Methods

We performed a retrospective cohort study of 12 consecutive adult patients with a prior history of congenital heart disease who underwent orthotopic transplantation at a single, large, academic center between September 1, 2005, and September 1, 2013.

Patient Population

The demographic data collected included age, weight, gender, and ethnicity. The preoperative data collected

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included congenital diagnosis and current anatomy, medical comorbidities, and condition at the time of transplantation. The operative data collected included type of surgeon, location of operation, number of prior operations, and duration of cardiopulmonary bypass and operation. The postoperative data collected included the need for mechanical support or tracheostomy, adverse events, the need for noncardiac consultants, and survival status. All adverse events were identified, in addition to those typically reported in the Society of Thoracic Surgeons database (operative mortality, stroke, renal failure, prolonged ventilation, deep sternal infection, reoperation, length of stay >6 days, and length of stay >14 days).

Care Environment

All of the operations were performed by a surgeon specializing in congenital heart disease in an adult hospital. Postoperative care was provided jointly by that surgeon and the adult cardiac transplantation team, also within the adult hospital. Long-term follow-up was provided by the adult cardiac transplantation team in the outpatient clinic.

For comparison, we queried other programs treating adults with congenital heart disease (n = 11) in the United States to assess the environmental variability in which transplantations in these adults are performed. The programs queried included those whose programs for adults with congenital cardiac disease are represented in the Alliance for Adult Research in Congenital Cardiology (AARCC). The programs were University of Michigan, Ann Arbor; Brigham and Women's Hospital/Harvard Medical School, Boston; Nationwide Children's Hospital/Ohio State University, Columbus; Heart Institute/Children's Hospital of Pittsburgh of UPMC, Pittsburgh; Hershey Medical Center/Pennsylvania State University, Hershey; Medical College of Wisconsin, Milwaukee; University of Colorado, Denver; Knight Cardiovascular Institute/Oregon Health and Science University, Portland; Texas Children's Hospital/Baylor College of Medicine, Houston; Children's National Medical Center, Washington, DC; and University of California, Los Angeles.

Institutional Review Board

Institutional review board approval was obtained to perform this retrospective study, and individual patient consent was waived. A descriptive analysis is reported throughout.

Results

Characteristics of Patients

At transplantation, the patients' median age and weight were 41 years (range, 16 to 72 years) and 65 kg (range, 45 to 104 kg) (Table 1); 58% were male, 92% were white, and 8% were African-American. The original cardiac diagnoses included tricuspid atresia (3), double-inlet left ventricle (2), unbalanced atrioventricular septal defect (2), congenitally corrected transposition of the great arteries (ccTGA) (2), and D-transposition of the great arteries (D-TGA) (3). The 7 patients with single-ventricle heart disease were listed for transplantation secondary to Fontan failure. The remaining 5 patients, 2 with CCTGA and 3 with D-TGA after atrial switch operations, were listed for transplantation secondary to systemic right ventricular failure. Six patients were called in from home, and 6 patients were in the hospital at the time of transplantation. The median time from admission to operation was 4 days (range, 0 to 148 days). One patient was intubated while in cardiogenic shock, and 5 patients were receiving support

Table 1. Demographics and Preoperative Characteristics

	Total Patients (n = 12)			
Characteristic	n	%	Median	Range
Demographics				
Age at operation (days)			41	(20–47)
Weight at operation (kg)			65	(45–104)
Gender				
Male	7	58	…	…
Female	5	42	…	…
Race				
White	11	92	…	…
African-American	1	8	…	…
Other	0	0	…	…
Congenital diagnosis/current anatomy				
Single ventricle - s/p Fontan operation	7	58	…	…
ccTGA	2	17	…	…
D-TGA - s/p atrial switch operation	3	25	…	…
Preoperative medical history				
Peripheral vascular disease	0	0	…	…
Cerebrovascular disease	5	42	…	…
Chronic lung disease	1	8	…	…
Diabetes	0	0	…	…
Hypertension	3	25	…	…
Endocarditis	0	0	…	…
Immunosuppressive treatment	2	17	…	…
Renal failure - dialysis	2	17	…	…
Hyperlipidemia	2	17	…	…
Tobacco	0	0	…	…
Preoperative cardiac status				
UNOS status				
IA	3	25	…	…
IB	9	75	…	…
NYHA class 4	12	100	…	…
Intubated	1	8	…	…
Cardiogenic shock	1	8	…	…
Inotropes	5	42	…	…
Mechanical support	0	0	…	…

ccTGA = congenitally corrected transposition of the great arteries; D-TGA = transposition of the great arteries; NYHA = New York Heart Association; s/p = status post; UNOS = United Network for Organ Sharing.