Non-Fontan Adult Congenital Heart Disease Transplantation Survival Is Equivalent to Acquired Heart Disease Transplantation Survival

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Background. As a result of improved diagnostic methods, medical treatment, surgical correction, and palliation in childhood, there is a growing number of adult patients with congenital heart disease (CHD) who may experience heart failure and subsequently require heart transplantation (HT). Because of complex anatomy, previous operations, and frequently increased pulmonary vascular resistance (PVR), these patients represent a group with a higher risk of early mortality after transplantation.

Methods. From May 1999 to December 2014, our institution performed 25 HTs in adult patients with end-stage CHD. We present our data and outcomes of transplantation in this group.

Results. The median age at transplantation was 38 years (range, 18.4–53.7 years). Survival was 88% at 30 days, 88% at 1 year, and 77% at 5 years. We identified long donor heart ischemic time (>4 hours) as an important risk factor for early mortality. There was no significant difference in the survival of patients undergoing transplantation for CHD and patients undergoing transplantation for other diagnoses.

Conclusions. With careful donor and recipient selection, adults with end-stage CHD undergoing HT can achieve excellent early and midterm survival, comparable to the survival of patients who undergo transplantation for other diagnoses.

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Congenital heart disease (CHD) is one of the most frequent congenital malformations. The incidence of moderate and severe forms of CHD is about 6 per 1,000 live births [1]. Advances in medical and surgical management have improved the long-term survival of patients with CHD [2]. Although in the 1950s only about 20% of newborns with moderate or complex CHD survived their first year, today up to 90% of these children live to adulthood [3]. However, despite the improving range of treatment options, many of these patients experience severe cardiac dysfunction in adult life. Heart transplantation (HT) or combined heart-lung transplantation may be the only choice for a growing number of adults with CHD and end-stage heart failure [4, 5]. In this study we review our results of HTs in adult CHD (ACHD), evaluate short- and midterm survival outcomes, and identify risk factors for mortality.

Patients and Methods

The Institute for Clinical and Experimental Medicine is 1 of 2 heart transplant centers in the Czech Republic. The adult cardiac transplantation program started in 1984. From January 1984 to December 2014 a total of 985 HTs were performed. The first patient with ACHD underwent transplantation in May 1999. Using our transplant database, we identified another 26 adult patients diagnosed with CHD who underwent heart or heart-lung transplantation from May 1999 until December 2014. Two patients who underwent combined heart-lung transplantation were excluded from this study. Thus a total of 25 patients with ACHD underwent isolated HT from 1999 until 2014.

Patient data were obtained from our transplantation database and hospital medical records. Demographic data, pretransplantation diagnoses, previous operations, pulmonary vascular resistance (PVR), clinical data, posttransplantation complications, and survival were reviewed. We compared posttransplantation survival of patients with ACHD with survival of 634 patients with other diagnoses who underwent HT in the same period (1999–2014).

We used the same protocol for HT in patients with CHD and patients with other diagnoses. In standard practice we use 1,000 to 1,500 mL of St. Thomas Hospital cardioplegic solution when harvesting the heart. The retrieval process itself is modified only by an effort to maximize the length of the donor vessels and make it possible to correct potential structural or positional abnormalities. After performing a median sternotomy in the recipient, extracorporeal circulation is established by

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cannulation of the ascending aorta and both venae cavae; however, if the preoperative tomographic scan indicates an intimate relationship between the aorta or right ventricle and the sternum, we insert the arterial cannula for extracorporeal circulation from the groin. After removing the donor heart from the transport container, we apply cold blood cardioplegia, which we repeat every 20 minutes during the anastomosis construction. Depending on the seriousness of perioperative bleeding, we either perform a primary closure of the sternotomy or delay closure of the sternotomy until blood coagulation measurements have normalized.

Immunosuppressive prophylaxis at our center includes preoperative and early postoperative administration of polyclonal antithymocyte globulin and a combination of calcineurin inhibitors (currently we predominantly use tacrolimus). Mycophenolate mofetil (MMF) and prednisone are given as maintenance therapy.

Demographic and clinical data are presented as the median, with the range of values in parentheses. Fisher's exact test was used to identify risk factors for 30-day, 1-year, and 5-year mortality. Risk factors included donor heart ischemic time longer than 4 hours, recipient PVR greater than 4 Wood units (WU), single-ventricle physiology, history of previous surgical intervention, and ventricular assist device (VAD) implantation in the recipient. The Mann-Whitney test was used for comparison of donor heart ischemic time between the 2 groups. Long-term survival was evaluated using Kaplan-Meier survival curves. Kaplan-Meier survival analysis and the log-rank test were performed to compare postoperative survival between the groups.

Results

A total of 25 adult patients with end-stage CHD (17 men [68%] and 8 women [32%]) underwent cardiac transplantation in our institution from 1999 until 2014.

Diagnoses, previous surgical intervention, PVR, donor heart ischemic time, and outcomes after transplantation are shown in chronologic order by date of transplantation in Table 1. The median age at transplantation was

Table 1. Pretransplantation Diagnoses, Previous Surgical Interventions, PVR, Donor Heart Ischemic Time, and Outcome After HT[a]

Patient	Age (years)	Diagnosis	Previous Operation	PVR	Ischemic Time	Survival	Status
1	20	DORV, VSD	PA banding, Kawashima procedure	1.8	116	15.6 y	Alive
2	42	Tetralogy of Fallot	BT shunt, repair of TOF	0.9	195	14.8 y	Alive
3	18	TGA	Mustard procedure	3.9	260	1 d	Dead
4	29	DOLV, PS	Septation of single ventricle	1.5	185	8.7 y	Dead
5	46	TGA	Mustard procedure	2.7	125	12.7 y	Alive
6	39	DILV, TGA, PS	None	3.0	220	12.3 y	Alive
7	50	DILV, TGA, PS	None	0.9	115	4.7 y	Dead
8	54	ALVT	Closure of the ALVT	1.4	240	9.6 y	Alive
9	52	Ebstein's anomaly	None	1.8	170	9.6 y	Alive
10	42	DILV, VSD, ASD	PA band, Ao-PA shunt, atrial septectomy	4.2	138	7.8 y	Alive
11	51	Tetralogy of Fallot	Repair of TOF	1.9	110	24 d	Dead
12	38	DORV, TGA, VSD, PS	Ao-PA shunt, atrial septectomy	2.4	100	7.1 y	Alive
13	30	TGA	Mustard procedure	0.6	177	7.0 y	Alive
14	23	TGA	Senning procedure	0.5	202	1.7 y	Dead
15	47	CC-TGA	None	2.6	98	6.5 y	Alive
16	30	TGA	Mustard procedure, LVAD HM II	1.2	75	5.6 y	Alive
17	35	VSD, CA, PLSVC	VSD closure, AVR, LVAD HM II	1.1	140	4.9 y	Alive
18	46	Ebstein's anomaly	TVP, RV plication, ASD closure	0.8	193	4.5 y	Alive
19	32	TGA	Mustard procedure, LVAD HM II	2.4	300	0 d	Dead
20	20	Pulmonary atresia, VSD	Ao-PA shunt, Rastelli procedure, PA stent	1.2	170	2.7 y	Alive
21	34	TGA	Mustard, LVAD HM II	1.8	117	2.3 y	Alive
22	49	Tetralogy of Fallot	BT shunt, repair of TOF	3.4	80	0.9 y	Alive
23	28	TGA, VSD	Senning procedure	1.4	90	0.7 y	Alive
24	49	Tetralogy of Fallot	BT shunt, repair of TOF	2.6	75	0.5 y	Alive
25	30	DORV, VSD, ASD, PS	LV-Ao connection, VSD + ASD repair, TVR	0.9	165	0.5 y	Alive

[a] In chronologic order by date of transplantation.

ALVT = aortico-left ventricular tunnel; Ao-PA shunt = aortopulmonary shunt; ASD = atrial septal defect; AVR = aortic valve replacement; BT shunt = Blalock-Taussig shunt; CA = coarctation of the aorta; CC-TGA = congenitally corrected transposition of the great arteries; DILV = double-inlet left ventricle; DOLV = double-outlet left ventricle; DORV = double-outlet right ventricle; HM II = Heart Mate II; HRV = hypoplastic right ventricle; HT = heart transplantation; LVAD = left ventricular assist device; LV-Ao = left ventricle to aorta; PA = pulmonary artery; PLSVC = persistent left superior vena cava; PS = pulmonary stenosis; PVR = pulmonary vascular resistance; RV = right ventricular; TGA = transposition of the great arteries; TOF = tetralogy of Fallot; TVP = tricuspid valve repair; TVR = tricuspid valve replacement; VSD = ventricular septal defect.