JACC Vol. 26, No. 3
September 1995:743–9

Heart Transplantation in Children With Congenital Heart Disease

DAPHNE T. HSU, MD, FACC, JAN M. QUAEGEBEUR, MD, ROBERT E. MICHLER, MD, FACC, CRAIG R. SMITH, MD, FACC, ERIC A. ROSE, MD, FACC, MARYANNE R. KICHUK, MD, WELTON M. GERSONY, MD, FACC, JUDITH F. DOUGLAS, RN, LINDA J. ADDONIZIO, MD, FACC

New York, New York

Objectives. **The aim of this study was to describe heart transplantation in children with congenital heart disease and to compare the results with those in children undergoing transplantation for other cardiac diseases.**

Background. **Reports describe decreased survival after heart transplantation in children with congenital heart disease compared with those with cardiomyopathy. However, transplantation is increasingly being considered in the surgical management of children with complex congenital heart disease. Present-day results from this group require reassessment.**

Methods. **The diagnoses, previous operations and indications for transplantation were characterized in children with congenital heart disease. Pretransplant course, graft ischemia time, posttransplant survival and outcome (rejection frequency, infection rate, length of hospital stay) were compared with those in children undergoing transplantation for other reasons (n = 47).**

Results. **Thirty-seven children (mean [±SD] age 9 ± 6 years) with congenital heart disease underwent transplantation; 86% had undergone one or more previous operations. Repair of extracardiac defects at transplantation was necessary in 23 patients. Causes of death after transplantation were donor failure in two patients, surgical bleeding in two, pulmonary hemorrhage in one, infection in four, rejection in three and graft atherosclerosis in one. No difference in 1- and 5-year survival rates (70% vs. 77% and 64% vs. 65%, respectively), rejection frequency or length of hospital stay was seen between children with and without congenital heart disease. Cardiopulmonary bypass and donor ischemia time were significantly longer in patients with congenital heart disease. Serious infections were more common in children with than without congenital heart disease (13 of 37 vs. 6 of 47, respectively, p = 0.01).**

Conclusions. **Despite the more complex cardiac surgery required at implantation and longer donor ischemic time, heart transplantation can be performed in children with complex congenital heart disease with success similar to that in patients with other cardiac diseases.**

(J Am Coll Cardiol 1995;26:743–9)

Surgical advances have greatly increased the survival of children with congenital heart disease. However, myocardial failure can occur early or late after operation for complex congenital heart disease. Late cardiac mortality has been reported after surgical repair of tetralogy of Fallot (5%), aortic stenosis (17%) and the Mustard procedure for transposition of the great arteries (15%) (1). Fontan et al. (2) reported an increase in risk of death from 6 to 15 years after the Fontan operation.

As survival after heart transplantation has improved, this option is more often considered in the surgical management of children with complex congenital heart disease. According to the registry of the International Society for Heart Transplantation (3), congenital heart disease has become the primary indication for transplantation in the pediatric population, with the proportion of children with congenital heart disease undergoing transplantation increasing from 16% in 1984 to 46% in 1993. Beyond the neonatal age range, most children with congenital heart disease undergoing transplantation have undergone one or more attempts at palliative or corrective cardiac surgery. Heart transplantation in these children presents a number of special problems. The indications and optimal timing of transplantation can be difficult to determine in patients with complex heart disease, and the type of reconstructive surgery of extracardiac defects required at transplantation must be carefully considered. Previous studies have reported many unique technical approaches to heart transplantation in patients with systemic and pulmonary venous anomalies (4–6), pulmonary artery stenoses (7), hypoplastic left heart syndrome (8), dextrocardia (9) and transposition of the great arteries (10–12).

It has been reported (13,14) that mortality among children with congenital heart disease is higher than that for children undergoing transplantation for other reasons, but recent studies (15,16) have shown improved survival. The present study describes the pretransplantation and posttransplantation courses of a relatively large series of children with and without

From the Departments of Pediatrics and Surgery, College of Physicians and Surgeons, Columbia University, New York, New York.

Manuscript received March 25, 1994; revised manuscript received December 27, 1994, accepted May 1, 1995.

Address for correspondence: Dr. Daphne T. Hsu, College of Physicians and Surgeons, Columbia University, Babies Hospital, 2 North, 3959 Broadway, New York, New York 10032.

0735-1097/95/$9 50
0735-1097(95)00253-7

Table 1. Diagnosis and Surgical History for 37 Study Patients

Pt No.	Diagnosis	Age (yr)	Surv (mo)	Status	Previous Operation
1	Hypoplastic left heart syndrome	0.0	0.5	Dead	None
2	Ebstein's anomaly	0.1	0.3	Dead	None
3	Single right ventricle, common atrium, *d*-malposition of the great arteries, pulmonary stenosis, asplenia syndrome	7.3	49.4	Alive	None
4	VSD, cardiomyopathy	12.7	55.6	Alive	None
5	Ventricular inversion, *l*-transposition of the great arteries, tricuspid insufficiency, complete heart block	18.1	61.8	Alive	None
6	Pulmonary atresia, intact ventricular septum, coronary artery fistulae	0.5	31.4	Alive	Ao-PA shunt
7	Single right ventricle, complete AV canal defect, mitral atresia, *d*-transposition of the great arteries, pulmonary stenosis, anomalous systemic venous return, polysplenia syndrome	1.1	26.0	Alive	Ao-PA shunt
8	Double-outlet right ventricle, complete AV canal defect, hypoplastic mitral valve, pulmonary stenosis, aortic stenosis	1.2	0.9	Dead	Ao-PA shunt
9	AV canal defect (complete), pulmonary stenosis	2.1	4.7	Dead	Ao-PA shunt
10	Double-outlet right ventricle, *d*-malposition of the great arteries, VSD, hypoplastic mitral valve, pulmonary atresia	8.0	39.4	Alive	Ao-PA shunt
11	Tricuspid atresia, small VSD, pulmonary atresia, mitral insufficiency	17.7	90.8	Alive	Ao-PA shunt
12	*d*-Transposition of the great arteries, VSD, mitral atresia, pulmonary stenosis	4.6	123.5	Alive	PA band; atrial septectomy
13	AV canal defect (partial), subaortic stenosis	12.8	25.6	Alive	Repair; Konno procedure
14	*d*-Transposition of the great arteries, VSD, coarctation of the Ao	13.4	40.7	Alive	Coarctation repair; Mustard procedure and VSD closure
15	Hypertrophic cardiomyopathy, coarctation of the Ao	15.8	77.8	Alive	Coarctation repair; myectomy
16	Shone's complex, coarctation of the Ao, aortic stenosis, subaortic stenosis	12.3	35.2	Alive	Coarctation repair; aortic valvotomy; aortic valve replacement; Konno procedure
17	Shone's complex, coarctation of the Ao, aortic stenosis, subaortic stenosis	3.7	46.1	Alive	Coarctation repair; aortic valvotomy; Konno procedure
18	Tetralogy of Fallot	13.8	92.4	Dead	Repair
19	Tetralogy of Fallot, pulmonary atresia	12.7	0.0	Dead	Repair; conduit replacement
20	Tetralogy of Fallot, tricuspid insufficiency	3.7	17.0	Alive	Repair; tricuspid valve annuloplasty
21	Ventricular inversion, *l*-transposition of the great arteries, VSD, pulmonary atresia, severe tricuspid insufficiency	3.7	54.2	Alive	Ao-PA shunt; repair
22	Ventricular inversion, *l*-transposition of the great arteries, VSD, pulmonary stenosis, dextrocardia, tricuspid insufficiency	13.8	39.9	Dead	Repair; tricuspid valve replacement
23	Ventricular inversion, *l*-transposition of the great arteries, VSD, pulmonary stenosis, severe tricuspid insufficiency	16.8	0.0	Dead	Repair; tricuspid valve replacement
24	Ventricular inversion, *l*-transposition of the great arteries, VSD, subaortic obstruction	18.3	26.0	Alive	PA band; VSD closure
25	Multiple VSDs	4.5	1.6	Dead	Repair
26	Double-inlet LV, ventricular inversion, *l*-malposition of the great arteries	15.9	60.3	Alive	PA band; Fontan
27	Double-inlet LV, ventricular inversion, *l*-malposition of the great arteries, restrictive bulboventricular foramen	15.1	0.9	Dead	PA Band; Fontan; Fontan revision; LV-Ao conduit placement
28	Double-inlet LV, ventricular inversion, *l*-malposition of the great arteries, pulmonary stenosis	11.9	50.2	Alive	Ao-PA shunts; Fontan
29	Double-inlet LV, ventricular inversion, normally related great arteries, bulboventricular foramen stenosis	2.3	26.2	Alive	Ao-PA shunt; bidirectional Glenn procedure
30	Double-inlet LV, ventricular inversion, normally related great arteries, pulmonary stenosis	15.3	29.1	Alive	Ao-PA shunt; Glenn shunt; Fontan
31	Double-outlet hypoplastic right ventricle, tricuspid stenosis, pulmonary stenosis, VSD, *l*-malposition of the great arteries	17.0	19.3	Alive	Fontan
32	Single LV, straddling tricuspid valve, hypoplastic right ventricle, *d*-malposition of the great arteries	15.0	0.0	Dead	Ao-PA shunts; Fontan; Fontan revision; Fontan takedown
33	Single LV, ventricular inversion, *l*-malposition of the great arteries, left AV valve atresia, bulboventricular stenosis	3.8	0.0	Dead	PA band, atrial septectomy; bulboventricular foramen enlargement; Fontan
34	Tricuspid atresia, VSD, hypoplastic right ventricle, pulmonary stenosis, normally related great arteries	10.3	20.1	Alive	Ao-PA shunt; Fontan
35	Tricuspid atresia, VSD, pulmonary stenosis, normally related great arteries	18.2	14.6	Alive	Ao-PA shunt; Glenn shunt; Fontan
36	Hypoplastic left heart syndrome	1.2	9.9	Dead	Norwood 1
37	Hypoplastic left heart syndrome	0.9	24.7	Alive	Norwood 1

Ao = aorta; AV = atrioventricular; PA = pulmonary artery; Pt = patient; LV = left ventricle; Surv = survival; VSD = ventricular septal defect.

Table 2. Indications for Transplantation in 37 Children With Congenital Heart Disease

Indication	No. of Pts
Reparative operation not feasible	2
Ventricular failure (no previous cardiac surgery)	3
Ventricular failure (after cardiac surgery)	23
Surgical repair	13
Palliative operation	8
Norwood 1 procedure	2
Low cardiac output after Fontan operation	9

Pts = patients.

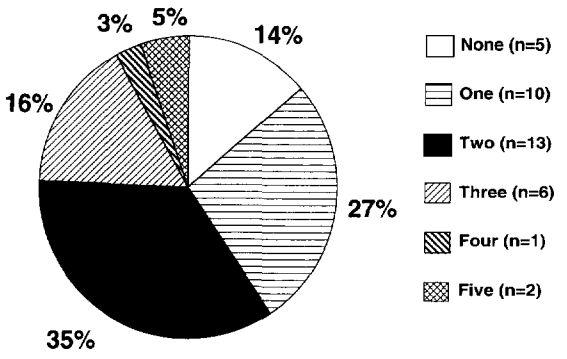

Figure 1. Number of cardiac surgery procedures per patient performed before transplantation in 37 patients.

congenital heart disease undergoing transplantation and compares their outcome.

Methods

Patients. Eighty-four consecutive children undergoing primary heart transplantation from June 1984 to August 1993 were included in the present study (44 boys, 40 girls; mean [±SD] age at time of transplantation 9.3 ± 6.2 years, range 1 week to 18.5 years; mean follow-up time from transplantation 37 ± 31 months, range of 0.1 to 122). The follow-up period ended in July 1994. Two patients underwent heterotopic heart transplantation because of severely elevated pulmonary vascular resistance, and 82 underwent orthotopic transplantation.

Indication for heart transplantation. Congenital heart disease with intractable congestive heart failure or complex congenital heart disease with severe deterioration not amenable to surgical correction were the indications for transplantation in 37 patients (44%). Thirty-four patients were older than 6 months of age at the time of operation. The number of children per year with the diagnosis of congenital heart disease undergoing transplantation at our institution increased from 2 patients in 1984 to 10 in 1992. For the 47 patients without congenital heart disease, the indication for transplantation was congestive heart failure refractory to maximal medical therapy. The underlying diagnoses of these patients were dilated cardiomyopathy (27 patients), endocardiofibroelastosis (7 patients), hypertrophic cardiomyopathy (7 patients) and adriamycin-induced cardiomyopathy (5 patients). One additional patient underwent transplantation because of congestive heart failure and ventricular arrhythmias secondary to an unresectable left ventricular tumor.

Study groups. The patients were classified into two groups for comparison; those with congenital heart disease and those with other cardiac diseases. The following preoperative variables were compared between the two groups: gender, age at transplantation, duration of congestive heart failure before transplantation, number of inotropic medications required and pulmonary vascular resistance index before transplantation. The posttransplantation variables of donor heart ischemia time, cardiopulmonary bypass time, patient survival, duration of posttransplantation hospital stay, rejections per patient-month and serious bacterial or fungal infections requiring intravenous antibiotic agents in the early postoperative (<3 months) period were also compared between the two groups. The incidence of graft atherosclerosis and lymphoproliferative disorders was also compared.

Immunosuppression protocol. Before 1986, all children undergoing transplantation (n = 14) were maintained on cyclosporine (serum trough levels of 150 to 200 ng/dl) and prednisone (0.2 mg/kg body weight per day). Six of these children had congenital heart disease, and eight had cardiomyopathy. After November 1986, azathioprine was added to the protocol in doses of 1.5 to 2 mg/kg per day, and cyclosporine trough levels were maintained at 200 ng/dl (n = 70). Endomyocardial biopsies were performed on a routine basis, and histologic evidence of acute rejection was treated with increased corticosteroid doses. Refractory episodes of rejection were treated with OKT3, rabbit ATG, methotrexate or total lymphoid irradiation, as clinically indicated.

Statistical analysis. Continuous variables were compared between patients with and without congenital heart disease by unpaired *t* test. Chi-square analysis was used to compare discrete variables. Survival between the two groups was compared using the Wilcoxon and log-rank tests for the association of response with covariates. The power of the study to detect a 10% difference in survival was 30%.

Results

Patients with congenital heart disease. *Diagnoses, surgical histories and indications for transplantation.* The mean (±SD) age of the 37 children with congenital heart disease who underwent transplantation was 9.3 ± 6.4 years with a mean length of follow-up of 31.2 ± 29 months. The diagnoses and surgical histories of these children are presented in Table 1.

Five patients had not undergone cardiac surgery before transplantation. Reparative surgery was not feasible in two newborns; three patients developed severe ventricular dysfunction in addition to their congenital heart defect (Table 2). Thirty-two patients (86%) developed significant ventricular failure after palliative or corrective surgery. Eight patients were in New York Heart Association functional class III at the time of transplantation; 29 were in class IV. Twenty-six (70%)

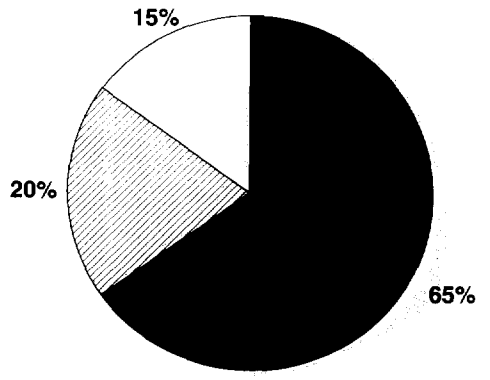

Figure 2. Closed heart procedures performed before transplantation in 20 patients: aorta to pulmonary artery shunt (n = 13, **solid area**); coarctation of the aorta (n = 4, **hatched area**); pulmonary artery banding (n = 3, **open area**).

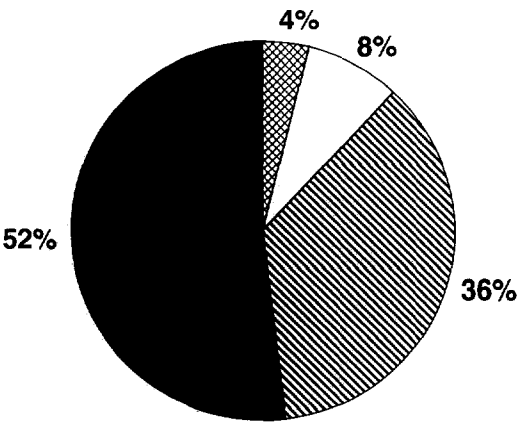

Figure 3. Open heart procedures performed before transplantation in 26 patients: bidirectional Glenn procedure (n = 1, **crosshatched area**); Norwood 1 (n = 2, **open area**); Fontan procedure (n = 9, **hatched area**); complete repair (n = 13; **solid area**).

of the 37 patients were receiving a mean of 1.8 (range 1 to 5) inotropic agents at the time of transplantation; eight were mechanically ventilated.

Twenty-two patients (60%) had undergone two or more cardiac procedures, and five previous operations had been performed in each of two patients (Fig. 1). Closed heart procedures were performed before transplantation in 20 patients (54%); the most common procedure was an aortopulmonary shunt (Fig. 2). Heart transplantation was performed after a closed heart procedure alone in seven patients, in six after an aortopulmonary shunt and in one after pulmonary artery banding. The remaining 13 patients underwent one or more additional cardiac surgical procedures before transplantation.

Twenty-five patients (68%) had undergone a mean of 1.6 ± 0.7 open heart surgeries before heart transplantation. The mean interval from the last open heart procedure to transplantation was 3.4 ± 3.0 years. Thirteen patients had undergone one or more operations to correct their congenital heart defect; 12 patients with a single ventricle had undergone palliative open heart surgery (Fig. 3).

Additional surgical procedures performed at the time of transplantation included takedown of a Fontan or Glenn procedure, repair of pulmonary artery stenosis and rerouting of the systemic or pulmonary venous return (Table 3). In all patients who required reconstruction of the systemic venous return, endomyocardial biopsies could be performed, when necessary, through the rerouted superior or inferior vena cava.

Mortality and cause of death. Thirteen of the 37 patients with congenital heart disease died after transplantation, 11 (85%) within 9 months after the procedure. Causes of death included donor heart failure, right heart failure secondary to pulmonary hypertension, postoperative hemorrhage, pulmonary hemorrhage, infection, rejection and graft atherosclerosis (Table 4). Of the three late deaths, one resulted from pneumonia after a prolonged hospital stay, and two occurred in adolescents who died at 40 and 92 months posttransplantation because of noncompliance with their immunosuppression protocol.

Residual extracardiac defects. Four patients had the presence of aortopulmonary collateral channels documented by descending aorta angiography after transplantation. Three had undergone a Fontan procedure, and one had two previous Blalock–Taussig shunts placed for severe pulmonary stenosis in the context of a polysplenia syndrome. In three of the four patients, high output heart failure developed within 2 weeks of transplantation, necessitating increased inotropic support and coil embolization of the major collateral channels.

Functional status. Twenty-two of the 25 patients currently alive are participating fully in age-appropriate activities. One 11-year old patient is limited by severe scoliosis and restrictive lung disease and attends school half-time. Two preschool children are mildly hypotonic and are undergoing physical therapy for delayed gross motor function.

Comparison: Patients with and without congenital heart disease. *Preoperative variables.* The pretransplantation clinical profiles of the patients with and without congenital heart disease indicated no significant difference between the groups with respect to age at transplant, gender, duration of congestive heart failure or number of inotropic medications required before transplantation (Table 5). The mean pulmonary vascular resistance index was relatively high for both groups, but there was no significant difference between the two groups.

Patient survival. There was no significant difference in patient survival between the two groups (Fig. 4). Three patients with congenital heart disease have undergone retransplantation for ventricular failure secondary to chronic rejection; however, overall graft survival was also not significantly different between the two groups. Regardless of diagnosis, overall survival after transplantation was significantly lower in all patients undergoing transplantation before mid-1986 compared with those undergoing transplantation since mid-1986. Only 2 (14%) of 14 children who underwent transplantation before mid-1986 still survive compared with 53 (75%) of 70

Table 3. Additional Surgical Procedures in 23 Patients at Time of Transplantation

Procedure	No. of Pts
Takedown of Fontan or Glenn procedure	10
Repair of branch pulmonary artery stenosis	10
Intraatrial baffle of left-sided SVC and/or IVC to right atrium	2
Takedown Mustard procedure	1

IVC (SVC) = inferior (superior) vena cava; Pts = patients.

children who underwent transplantation since mid-1986. However, survival was not significantly different between patients with congenital heart disease and those with other cardiac diseases undergoing transplantation before mid-1986 (none of six vs. two of eight survivors). Similarly, no difference in survival was seen by diagnosis among patients undergoing transplantation after mid-1986. Ten of the 12 deaths that occurred in the patients who underwent transplantation before mid-1986 were due to rejection, whereas 3 of 17 deaths after mid-1986 were secondary to rejection. Thus, the type of immunosuppression was significantly associated with the difference in survival between the two time periods (17).

Intraoperative and postoperative variables. Intraoperative and postoperative variables were compared between the patients with and without congenital heart disease (Table 6). Graft ischemia time and cardiopulmonary bypass time were significantly longer in patients with congenital heart disease. The number of patients with an early bacterial or fungal infection after transplantation was significantly higher in the group with than without congenital heart disease (13 of 37 vs. 6 of 47, respectively). The infections seen in the group with congenital heart disease included bacterial sepsis (eight patients), bacterial and *Candida albicans* sepsis (two patients), *Staphylococcus aureus* and *Aspergillus* empyema (one patient), toxoplasmosis (one patient) and bacterial pneumonia (one patient).

Coronary artery disease and lymphoproliferative disease. The incidence of graft atherosclerosis was no different between children with and without congenital heart disease. Sixty of the 84 patients had selective coronary artery angiography or postmortem histologic examination of the coronary arteries of the transplanted heart. Five of 23 patients with and 5 of 37 patients without congenital heart disease had evidence of graft atherosclerosis.

Lymphoproliferative disorders have been diagnosed in 5 of the 84 patients in the present study. Three patients had the pretransplantation diagnosis of congenital heart disease, and two patients underwent transplantation because of end-stage congestive heart failure secondary to cardiomyopathy.

Table 4. Cause of Death in 13 Patients With Congenital Heart Disease

Cause of Death	No. of Pts
Donor failure	1
Right heart failure	1
Postoperative hemorrhage	2
Pulmonary hemorrhage	1
Infection (<6 mo after transplantation)	3
Aspiration pneumonia	1
Fungal and CMV sepsis	2
Acute rejection secondary to improper cyclosporine dosage	1
Graft atherosclerosis	1
Infection (9 mo after transplantation)	1
Noncompliance with immunosuppressive protocol	2

CMV = cytomegalovirus; Pts = patients.

Table 5. Comparison of Preoperative Variables

	CHD (n = 37)	No CHD (n = 47)	p Value
Age (yr)	9.4 ± 6.5	9.5 ± 6.1	NS
Gender (M/F)	22/15	32/15	NS
Duration of CHF (months)	15.2 ± 26.9	21.9 ± 28.5	NS
Inotropic support before transplantation (medications/pt)	1.2 ± 1.1	1.0 ± 1.0	NS
PVRI (U/m^2)	5.9 ± 3.4	6.8 ± 4.7	NS

Data presented are mean value ± SD or number of patients. CHD = congenital heart disease; CHF = congestive heart failure; pt = patient; PVRI = pulmonary vascular resistance index.

Discussion

Transplantation for congenital heart disease. The present study describes the use of transplantation as a surgical approach to children with congenital heart disease and end-stage heart failure. The majority of patients were outside the neonatal period and had undergone previous palliative or corrective surgery. Most patients had the clinical findings of severe ventricular dysfunction and were critically ill in the intensive care unit at the time of transplantation. Thus, expected 1-year survival in this patient group without transplantation was extremely low, in contrast to the 1-year survival of 70% and 5-year survival of 64% after transplantation.

Early studies suggested that congenital heart disease was an independent risk factor for death after heart transplantation in children outside the neonatal age range. Trento et al. (14) reported 4 survivors after transplantation in 10 patients with congenital heart disease; four of the six deaths were attributed to donor right heart failure. Right ventricular decompensation has also been reported as a cause of death in other series (18–20). Although the mean pulmonary vascular resistance index of the patients in the present study was 6.0 units, only one patient died of donor right heart failure. As previously reported from this institution (21), aggressive pretransplantation and posttransplantation measures to lower pulmonary vascular resistance and prevent donor right heart failure most likely contributed to the lower incidence of death from right heart failure. In a follow-up study by Armitage et al., 11

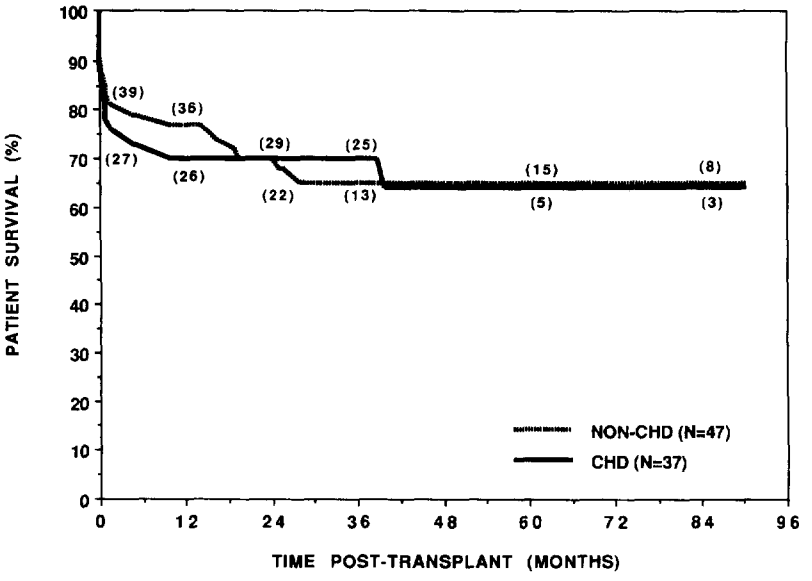

Figure 4. Patient survival after transplantation in patients with and without congenital heart disease (CHD).

additional patients with congenital heart disease were added to the original 10 patients, and the improved 1-year survival of 60% was attributed to "improved myocardial protection, surgical experience and more aggressive post-transplant care" (15).

Reported 1-year survival after transplantation in children with congenital heart disease ranges from 3 of 11 patients reported by Baum et al. (19) to 9 of 10 patients reported by Chartrand (18). The 70% 1-year survival rate in the present series is similar to that of 53% to 71% reported by other investigators (9,13,16). The majority (83%) of deaths in the present series occurred early after transplantation, although only two deaths from postoperative hemorrhage could be directly attributed to the complexity of the structural heart disease. The 5-year survival of 64% in the present study is also comparable to long-term survival after transplantation reported in the other smaller series (13,15,16).

Additional reconstructive surgery was performed at the time of transplantation in 62% of the patients in the present series. Many innovative techniques have been described (4–12) to reconstruct and repair extracardiac defects. However, to our knowledge comparison of donor ischemia time and cardiopulmonary bypass time between patients with and without congenital heart disease has not been reported previously. In the present study, both donor heart ischemia time and cardiopulmonary bypass time were longer in the patients with congenital heart disease, implying that the time required for reconstructive surgery before transplantation contributed to the duration of graft ischemia. Improved coordination of the timing of organ donor procurement to increase the operative time allotted to prepare the recipient with congenital heart disease for transplantation may help shorten donor ischemia time.

Table 6. Comparison of Intraoperative and Postoperative Variables

	CHD	No CHD	p Value
Donor heart ischemia time (min)	221 ± 70	180 ± 64	0.004
Cardiopulmonary bypass time (min)	220 ± 78	116 ± 37	0.0001
Posttransplantation hospital stay (days)	37 ± 51	28 ± 22	NS
Perioperative infection	13	6	0.01
Rejection/pt-mo	0.2 ± 0.57	0.15 ± 0.23	NS

Data presented are mean value ± SD or number of patients. CHD = congenital heart disease; pt = patient.

Infections. The incidence of significant postoperative infections in our patients with congenital heart disease was 35%. The most common infection was bacterial sepsis, although three patients did have fungal sepsis. Ten of the 13 patients had undergone previous cardiac surgery, which may have led to slower wound healing because of decreased vascularity of scar tissue and a predisposition to infection; however, the etiology of the increased susceptibility to infection in these patients is not clear.

Aortopulmonary collateral channels. The incidence of aortopulmonary collateral channels in patients is reported (22) to be 36% in patients after the Fontan or bidirectional Glenn procedure. Aortopulmonary collateral channels have not been reported after transplantation for congenital heart disease. The incidence in the present series was similarly 33% (3 of 9) in the patients after a Fontan procedure; two patients developed significant hemodynamic compromise and required catheter embolization of the collateral channels.

Summary. Transplantation should be considered under various circumstances for the child with complex congenital heart disease. The decision to replace rather than to attempt palliation or repair in very high risk cases must be tailored to the individual patient, weighing the complexity of the reparative surgery and the possibility of success against the risks of

the transplant operation and consequences of long-term immunosuppression in the pediatric patient. Some children have no other options because all other operative choices have been exhausted. At our institution, the only anatomic contraindication to heart transplantation is severe pulmonary artery hypoplasia or pulmonary vein stenosis. The present study demonstrates that despite longer graft ischemia and bypass times and a higher incidence of postoperative infections, survival after transplantation in children with congenital heart disease is comparable to survival after transplantation in children with other cardiac diseases, predominantly cardiomyopathy. Transplantation was performed successfully in six of nine patients with a "failed" Fontan procedure, offering these severely debilitated patients the possibility of a return to normal activities. In addition, six of nine patients with single-ventricle anatomy in whom a Fontan procedure was contraindicated also survived transplantation. Our data demonstrate that heart transplantation offers the possibility of excellent medium-term survival for children with a variety of complex congenital heart lesions and should be considered as a therapeutic option in children with myocar-dial dysfunction or in those patients with no other viable options.

References

1. Morris CD, Menashe VD. 25-Year mortality after surgical repair of congenital heart defect in childhood. JAMA 1991;266:3447–52.
2. Fontan F, Kirklin JW, Fernandez G, et al. Outcome after a "perfect" Fontan operation. Circulation 1990;81:1520–36.
3. Hosenpud JD, Novick RJ, Breen TJ, Daily OP. The registry of the International Society for Heart Transplantation: eleventh official report—1994. J Heart Lung Transplant 1994;13:561–70.
4. Doty DB, Renlund DG, Caputo GR, Burton NA, Jones KW. Cardiac transplantation in situs inversus. J Thorac Cardiovasc Surg 1990;99: 493–9.
5. Chartrand C, Guerin R, Kangah M, Stanley P. Pediatric heart transplantation: surgical considerations for congenital heart diseases. J Heart Transplant 1990;9:608–17.
6. Mayer JE, Perry S, O'Brien P, et al. Orthotopic heart transplantation for complex congenital heart disease. J Thorac Cardiovasc Surg 1990;99:484–91.
7. Cooper MM, Fuzesi L, Addonizio LJ, Hsu DT, Smith CR, Rose EA. Pediatric heart transplantation after operations involving the pulmonary arteries. J Thorac Cardiovasc Surg 1991;102:386–94.
8. Bailey LL, Wood M, Razzouk A, Van Arsdell G, Gundry S. Heart transplantation during the first 12 years of life. Arch Surg 1989;124:1221–6.
9. Menkis AH, McKenzie FN, Novick RJ, et al. Expanding applicability of transplantation after multiple prior palliative procedures. Ann Thorac Surg 1991;52:722–6.
10. Richenbacher WE, Karwande SV, Shaddy RE, et al. Cardiac transplantation in a patient with a single ventricle and transposition of the great vessels. Ann Thorac Surg 1991;52:871–3.
11. Harjula ALJ, Heikkila LJ, Neiminen MS, Kupari M, Keto P, Matilla SP. Heart transplantation in repaired transposition of the great arteries. Ann Thorac Surg 1988;46:611–4.
12. Rietz BA, Jamieson SW, Gaudiani VA. Method for cardiac transplantation in corrected transposition. J Cardiovasc Surg 1982;23:293–6.
13. Radley-Smith RC, Yacoub MH. Long-term results of pediatric heart transplantation. J Heart Lung Transplant 1992;11:S277–81.
14. Trento A, Griffith BP, Fricker FJ, Kormos RL, Armitage J, Hardesty RL. Lessons learned in pediatric heart transplantation. Ann Thorac Surg 1989; 48:617–23.
15. Armitage JM, Fricker FJ, del Nido P, Starzl TE, Hardesty RL, Griffith BP. A decade (1982 to 1992) of pediatric cardiac transplantation and the impact of FK 506 immunosuppression. J Thorac Cardiovasc Surg 1993; 105:464–72.
16. Hasan A, Au J, Hamilton JR, Hunter S, Hilton CJ, Dark JH. Orthotopic heart transplantation for congenital heart disease. Technical considerations. Eur J Cardiothorac Surg 1993;7:65–78.
17. Addonizio LJ, Hsu DT, Douglas JM, et al. Decreasing incidence of coronary disease in pediatric cardiac transplant recipients using increased immunosuppression. Circulation 1993;88 Suppl II (Pt 2):II224–39.
18. Chartrand C. Pediatric cardiac transplantation despite atrial and venous return anomalies. Ann Thorac Surg 1991;52:716–21.
19. Baum D, Bernstein D, Starnes VA, et al. Pediatric heart transplantation at Stanford: results of a 15-year experience. Pediatrics 1991;88:203–14.
20. Merrill WH, Frist WH, Stewart JR, et al. Heart transplantation in children. Ann Surg 1991;213:393–8.
21. Addonizio LJ, Hsu DT, Douglas JR, et al. Cardiac transplant in children with markedly elevated pulmonary vascular resistance [abstract]. J Heart Lung Transplant 1993;12:S93.
22. Triedman JK, Bridges ND, Mayer JE, Lock JE. Prevalence and risk factors for aortopulmonary collateral vessels after Fontan and bidirectional Glenn procedures. J Am Coll Cardiol 1993;22:207–15.