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Transplantation in complex congenital heart disease

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Abstract

Three hundred and forty-five pediatric patients underwent orthotopic heart transplantation at Loma Linda University Medical Center throughout August 1999. Seventy-five percent of these patients had the diagnosis of congenital heart disease as the primary indication for transplantation. Two hundred and fifty-seven were infants and of these, 91 were neonates. Forty-nine percent had the primary diagnosis of hypoplastic left heart syndrome or its equivalent. Thirty patients had variant situs and 15 of these had situs inversus. Seventy-seven children had undergone one or more previous cardiac and/or thoracic procedures. There appeared to be a survival advantage with neonatal as compared to infant heart transplants (77% vs. 63%). Technical considerations for reconstruction have made congenital heart transplantation a low perioperative risk factor as compared to the child with cardiomyopathy. Attention to (1) recipient selection; (2) adequate en bloc removal of the heart and accompanying vessels; (3) use of low-flow bypass techniques with limited circulatory rest; and (4) multidisciplinary management have been the fundamental approach at this institution. Although challenging, high perioperative survival and long-term success can be achieved with heart transplantation in patients with high-risk or inoperable congenital heart disease.

Keywords: Complex congenital heart defects; Hypoplastic left heart syndrome; Total anomalous pulmonary venous drainage; Situs inversus; Cardiopulmonary shunt; Low-flow suction bypass

1. Introduction

Currently, 74% of infant heart transplants are related to inoperable or high-risk congenital heart disease [1]. This reflects the most common indication for the pediatric population; although only 30% of transplanted children older than 1 year of age have this diagnosis. The 1999 report of the International Society for Heart and Lung Transplant Registry cites a 59% 6-year survival for infant heart transplantations. This figure is deceptive. The actuarial survival at Loma Linda University Medical Center for neonate and infant heart transplants is 77% and 63%, respectively, at 10 years. These figures reflect a possible salutary immunologic period for transplantation in the first month of life and further relates to the option of successful retransplantation ($n = 5$) in this same group.

The modern era of transplantation for neonatal congenital heart disease was introduced at Loma Linda University in Southern California. In 1985, Bailey et al. performed the first successful neonatal heart transplant for hypoplastic left heart syndrome. This child is alive and well at 14 years of age. The foundation of these techniques and those of Dr Adrian Kantrowitz, 16 years earlier, when he attempted a heart transplant on a neonate with Ebstein's anomaly, were built on experimental laboratory work with animals. Since this incipient work, heart transplantation has become available to infants and children with endstage congenital heart disease, as well as those infants and children affected by cardiomyopathic diseases.

It is the purpose of this report to outline the anatomic considerations in surgical treatment of con-

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genital heart disease and demonstrate the overall conglomerative surgical experience in enabling the high perioperative survival of this population (91%). Virtually all forms of endstage complex congenital heart disease are amenable to transplantation with the exception of atretic or absent pulmonary vasculature, and older infants and children with fixed pulmonary hypertensive disease (> 4 Wood units).

2. Patient population

Now through almost a decade and a half, Loma Linda has completed 345 orthotopic heart transplants in infants and children. Two hundred and fifty-seven were under 1 year of age and 97 of these were neonates. Forty-nine percent had the primary diagnosis of hypoplastic left heart syndrome or its equivalent. Of these, nine were associated with interrupted aortic arch and 12 with total anomalous pulmonary venous connection.

Twelve recipients had visceral heterotaxia. Seven had isolated dextrocardia, and 30 had variant atrial situs including 15 with situs inversus. Of those beyond neonatal age, 77 had been subjected to one or more previous cardiac or thoracic procedures. Nineteen recipients had undergone palliative procedures for establishment of univentricular physiology. Eight had the Norwood procedure and 14 had Glenn or Fontan cavopulmonary anastomoses.

There are four areas of technical consideration with regard to recipient reconstruction. This would involve: (1) aortic hypoplasia or interruption; (2) systemic venous anomalies; (3) situs variants; and (4) pulmonary vascular anomalies. As a result, en bloc removal of the donor heart could include extended veins, complete pulmonary arteries, and variable amounts of ascending aorta and aortic arch. To illustrate techniques required for successful orthotopic heart transplant among recipients with complex congenital cardiac anatomy, four different reconstruction patterns have been chosen: (1) hypoplastic left heart syndrome; (2) total anomalous pulmonary venous connection; (3) situs inversus; and (4) unilateral or bilateral cavopulmonary shunts with central pulmonary artery absence or stenosis.

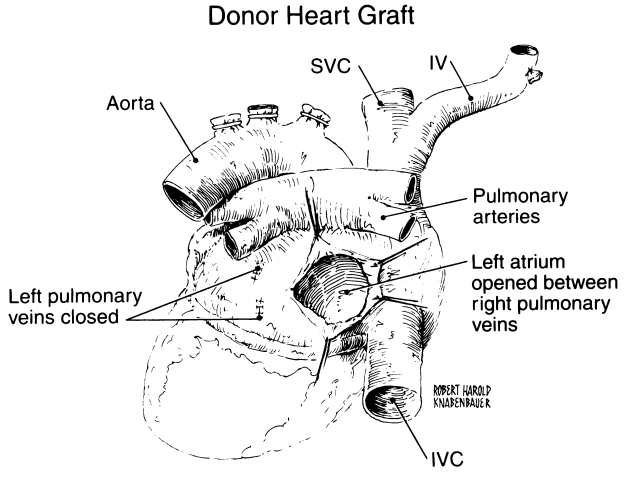

Fig. 1. Wide en bloc removal of the donor heart complete with both atria, extended systemic veins, complete central pulmonary arteries and variable amounts of ascending and arched aorta may be required for reconstruction.

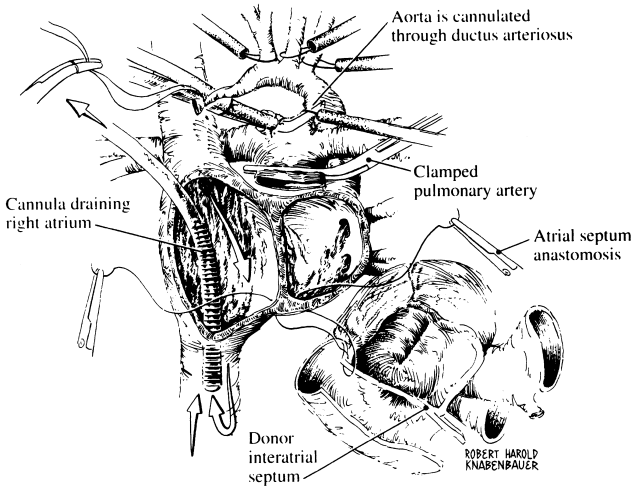

Fig. 2. With the pulmonary artery clamped, the atretic ascending aorta is ligated and divided while the recipient cardiectomy is performed. Hypothermic low flow suction bypass is used and the atrial anastomosis is begun at the inferior septum.

3. Donor heart procurement

Heart procurement relies on individual recipient anatomic requirements as outlined by the four technical considerations. Other organ procurement teams may affect the extent of en bloc resection of the heart. Specifically, concomitant lung procurement would preclude a heart recipient in need of central pulmonary artery replacement or one who has situs inversus or total anomalous pulmonary venous drainage. The latter two variations would require virtually all of the left atrium. Fig. 1 represents the appearance of the donor heart for transplantation in aortic arch and pulmonary artery reconstruction as well as recipients requiring an intact left atrium. Generally, tailoring of the donor specimen is withheld until implantation.

4. Recipient bypass

In infants and small children, a simple two-cannula

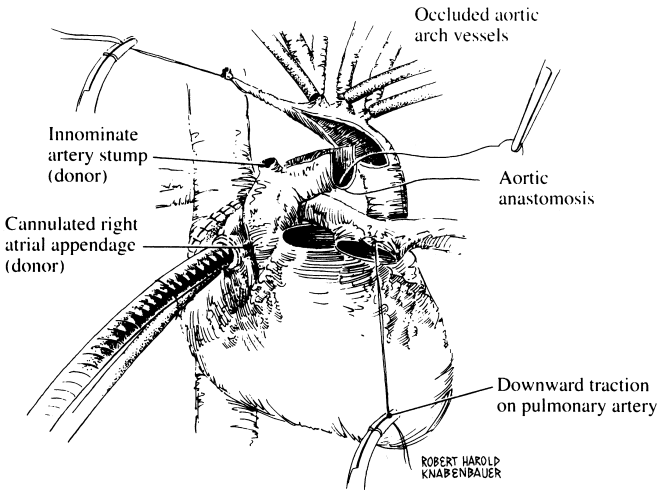

Fig. 3. Tourniquets placed around aortic arch vessels are tightened; ductus is ligated and divided. Under circulatory arrest, the under surface of the recipient arch is opened and reconstructed with donor arch aorta starting at the isthmus. The atrial venous cannula may be inserted along with the aortic cannula into the innominate artery stump to start reperfusion.

technique is used to initiate bypass. Profound hypothermia (20°C) is established and low-flow suction bypass is started once aortic separation occurs and cardiectomy has begun. Low-flow suction bypass (20–30 ml/kg/min) employs a flexible cannula which aspirates venous blood in the field and returns it to the oxygenator to be recycled to the patient. If aortic arch interruption is present, an equally split arterial perfusion system may be used.

The main theme in dealing with complex congenital heart disease has been to simplify the bypass methods to facilitate the operation. In recent years, low-flow hypothermic perfusion has lessened the circulatory arrest time. This has potentially decreased the possible neurologic sequelae of prolonged hypothermic circulatory arrest. Currently, median circulatory arrest times in hypoplastic left heart syndrome patients requiring aortic arch reconstruction has decreased from 53 ± 11 min to 26 ± 8 min [2].

4.1. Hypoplastic left heart syndrome

In this ductal dependent system, the aortic cannula is directed through the ductus from the main pulmonary artery and held in place by a silk tourniquet. With the venous cannula in the right atrium, core temperature is reduced to 18–20°C. Arch vessels are dissected and silk tourniquets are loosely applied. The atretic ascending aorta is ligated and divided with the distal stump ligature on loose traction. The left pericardium is extracted to allow extension of the donor heart into the left chest. The main pulmonary artery is clamped and low-flow suction bypass is inaugurated for cardiectomy (Fig. 2). The atrial septum and right

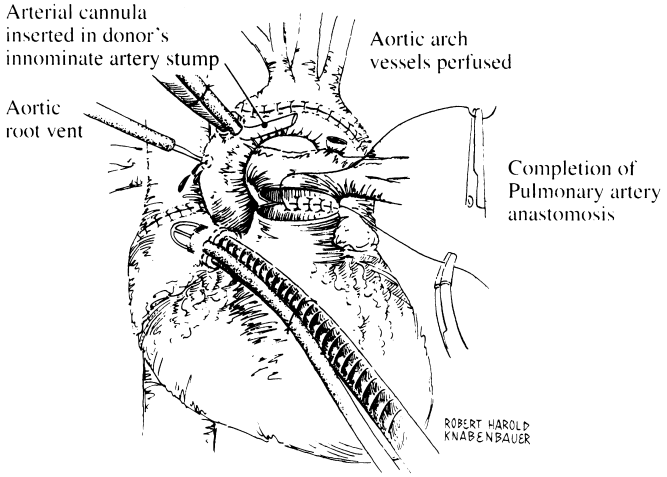

Fig. 4. With circulation restarted, air is removed by venting at the previous cardioplegia site in the donor aorta. The pulmonary artery anastomosis is performed while rewarming the patient.

atrial connections are accomplished first, followed by the left atrial anastomosis.

Circulatory arrest is begun once the arch vessel tourniquets are applied and the ductus is ligated and divided. With traction on the distal aortic stump ligature, the arch and aortic isthmus are exposed and opened. Excess ductal tissue is excised to prevent later coarctation. Arch reconstruction is begun by splaying the superior aspects of the donor aortic arch near to but not including the innominate artery stump (Fig. 3). This becomes the underside of the neoaortic arch. Before circulation is reestablished, normal saline is flushed through the donor innominate artery displacing air. This stump then receives the aortic cannula and bypass is restarted.

Shortly after this, arch vessel tourniquets are removed and systemic rewarming with graft reperfusion is begun. The donor-ascending aorta is vented through the previous cardioplegia cannulation site. The pulmonary arterial anastomosis is accomplished during reperfusion and can be done partially or completely with suction bypass (Fig. 4). At this point, standard venous cannulation and full bypass flows are reestablished. Graft reperfusion is continued for a minimum of 1 h to ensure donor heart functional recovery.

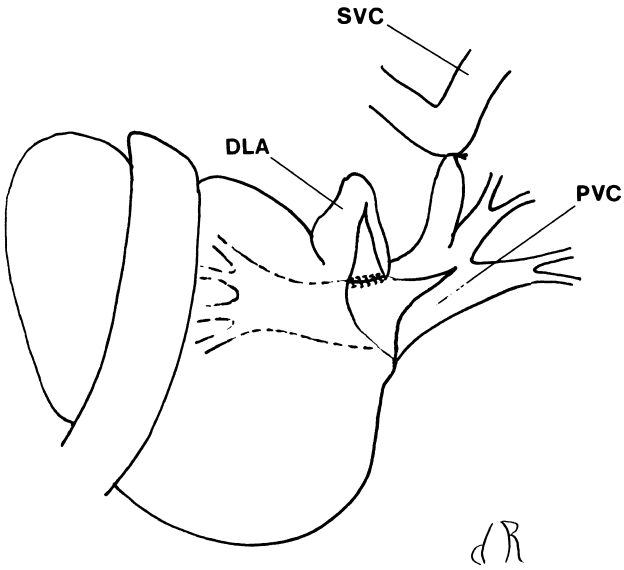

Fig. 5. This illustrates the technique of atrial anastomosis in a recipient with supra cardiac type of total anomalous pulmonary venous connection. A donor left atrial incision is created at the time of implantation and corresponds to the orientation of the common pulmonary venous channel. SVC, superior vena cava; PVC, common pulmonary venous channel; DLA, donor left atrium.

4.2. Total anomalous pulmonary venous connection with univentricular anatomy

Partial or complete total anomalous pulmonary venous connection may be part of univentricular malformations, especially in the setting of hypoplastic left heart syndrome. As in Fig. 1, the entire donor left atrium is removed by dividing the pulmonary veins individually. An opening is made in the left atrium connecting two or more pulmonary veins according to the orientation of the posterior common pulmonary venous confluence [3]. The openings can be transverse, oblique, or longitudinal depending on the type of anomalous pulmonary venous drainage pattern. The abnormal pulmonary venous connection to the systemic venous circulation is ligated and divided. Any additional donor pulmonary venous openings are closed. The anastomosis is achieved with running polypropylene (Fig. 5). The remainder of the implantation proceeds in a regular manner.

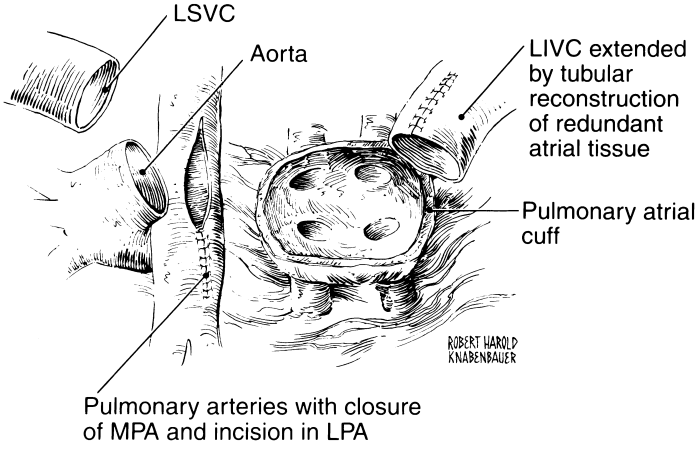

Fig. 6. After cardiectomy, the IV and attached portion of systemic atrium are constructed into a rightward directed conduit for donor IVC anastomosis. The pulmonary artery is opened to the left toward the LPA. LSVC, left superior vena cava; LIVC, left inferior vena cava; MPA, main pulmonary artery; LPA, left pulmonary artery.

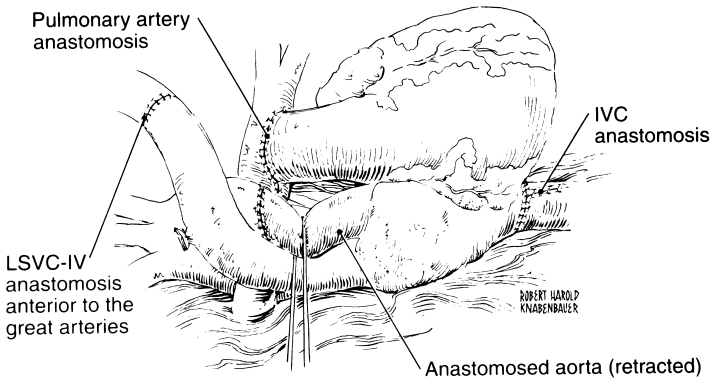

Fig. 7. The conduit formed by the donor's SVC and innominate vein is connected to the left-sided native superior vena cava, in front of the aortic and pulmonary anastomoses. LSVC-IV, left superior vena cava-innominate vein.

4.3. Situs inversus

Donor cardiectomy includes extended removal of the superior vena cava with the innominate vein and an intact left atrium. Donor left atrium is tailored by oversewing or ligating the left pulmonary veins, opening the space between the right pulmonary veins and extending this opening in a transverse plane as needed [4]. Since the recipient left atrium tends to be a midline structure, this helps to align the donor heart for the subsequent systemic venous and arterial connections. Bypass is inaugurated in the usual manner and profound hypothermia with low-flow bypass is employed. The left pericardium is excised for a leftward rotation of the graft.

Subsequent cardiectomy leaves a small left atrial cuff and a rim of atrial tissue on the inferior vena cava to be later fashioned into a rightward-directed conduit for the donor inferior vena cava (Fig. 6). The order of anastomosis proceeds with left atrial implantation and then inferior vena cava connection. Other techniques have been described for systemic venous reconstitution including spiral saphenous vein grafts [5], intraatrial baffles [6], and use of Dacron either as

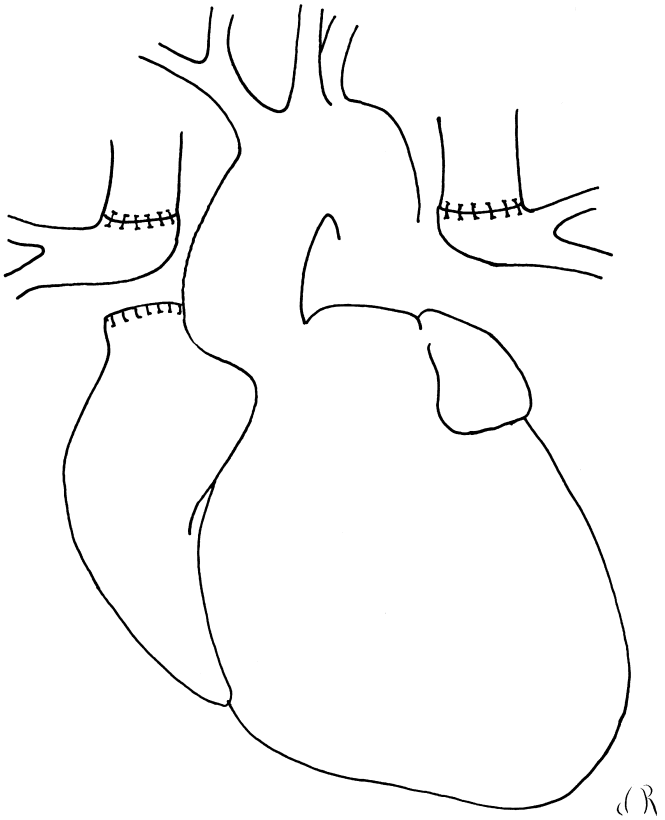

Fig. 8. Anatomy of a recipient subjected to bilateral bi-directional superior caval pulmonary shunts with absent central pulmonary arteries.

a septum or conduit [7,8]. The simple technique presented here relies on autologous and donor tissue, both of which possess growth potential.

Aortic anastomosis can follow, especially if arch reconstruction is necessary. This allows for earlier graft perfusion and placement of the donor superior vena cava-innominate vein conduit anteriorly. This configuration has been shown to be less likely to produce stenosis of the superior vena cava. The main pulmonary artery, which also tends to be a midline structure, is oversewn and a new opening is made extending towards the left pulmonary artery (Fig. 7). During graft reperfusion, the donor main pulmonary artery is then anastomosed to the left of the aorta.

4.4. Cavopulmonary shunts and stenosis or absence of main pulmonary artery

Graft implantation is further modified through native or created systemic venous and pulmonary artery anomalies. Cavopulmonary shunts and absent or stenotic pulmonary arteries are outcomes from the course of palliative surgery used to achieve stable univentricular physiology (Fig. 8). Standard cannulation and perfusion techniques are again employed and donor procurement requires en bloc extended systemic venous and complete pulmonary artery harvest. Recipient cardiectomy involves disconnection of previous cavopulmonary shunts and splaying of the stenotic main pulmonary artery or tailoring of bilateral pulmonary artery ends. Orientation of the graft is critical to avoid torsion. The sequence of implantation after left atrial connection is: (1) inferior cava; (2) right and left pulmonary arteries; (3) unilateral or bilateral superior vena cava; and (4) aorta (Fig. 9).

5. Discussion

Our experience confirms the effectiveness and safety of heart transplantation in patients with complex congenital heart disease with or without prior surgical intervention. This challenges the validity of congenital heart disease as a risk factor in pediatric transplantation. Numerous reports from this and other institutions show equivalent perioperative survival rates to the cardiomyopathic group [9–11]. It has also been shown that sequelae such as arteriovenous mal-

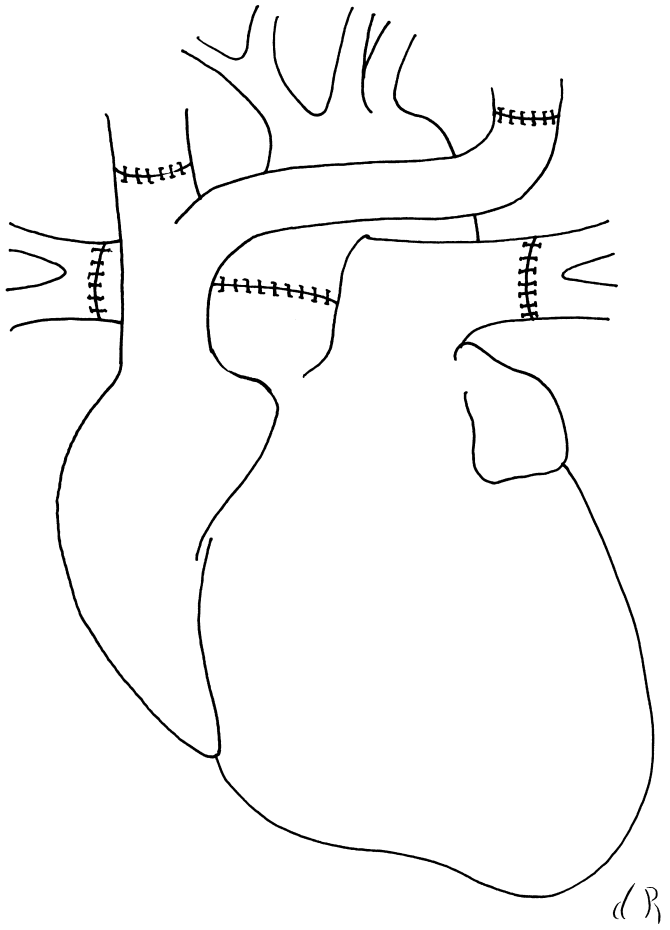

Fig. 9. The final appearance of the recipient after heart transplantation with reconstruction of systemic veins and bilateral pulmonary arteries.

formations created by cavopulmonary shunt physiology will resolve with transplantation [12]. Improvements in heart transplant survival are due to implementation of innovative surgical techniques, refined perioperative management for rejection and infection, and a multidisciplinary approach for long-term management and treatment of these children. It has been said that the donor shortage, which contributes to the 15–20% rate of death in the preoperative recipient population, converts transplantation for hypoplastic left heart syndrome to a modality of last resort [13]. In actuality, even with the recipient attrition, overall survival rates comparing major institutions with regard to transplantation vs. the Norwood procedure, are equivalent [14]. Certainly with respect to the trio of operations necessary to convert a hypoplastic left heart syndrome patient to univentricular physiology, there would appear to be a theoretical advantage if a single stage heart transplant operation could be performed.

The limiting factor will always be donor organ availability. With more than 100 deaths per day in the US among infants less than 1 year of age, it is hoped that news of this on-going need will reach the caring physician and the respective patient guardian. For now we look for alternate donor pools and a more specific enduring graft tolerance.

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