Heart Transplant: Transplantation for Congenital Heart Disease

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Cardiac transplantation for congenital heart disease incorporates aspects of both reparative and replacement surgery. Although intracardiac congenital malformations are replaced, and therefore pose few obstacles to the transplant surgeon, extracardiac malformations (congenital, acquired, or iatrogenic) can present considerable challenges to the operative team. Before transplantation, a full comprehension of the operative plan for the management of each individual patient is essential for both the donor team (so that they may harvest donor tissue of appropriate amounts to allow for adequate reconstruction) and the recipient anesthesi-surgical team (so that they may have several contingency plans for the safe establishment of cardiopulmonary bypass and full cardiac support during cardiectomy). Several recent publications have addressed quite extensively various surgical strategies aimed at heart transplantation for complex congenital heart disease; accordingly, the focus of this article is to address those more “common” anatomical challenges, some of which may be encountered in particular by noncongenital heart surgeons, given the increasing incidence of adult patients with congenital disease who have end-stage heart failure.1,2

For congenital cardiac transplant candidates, a standard, systematic approach toward surgical planning is recommended.3 Key considerations include issues of atrial situs, anomalies of systemic venous return, anomalies of the great arteries, and particular problems related to prior catheter-based palliations. Where possible, maximizing the “preparation” (reconstruction) of the recipient anatomy that can be accomplished before implant of the donor heart helps to reduce the overall warm ischemic time (and if well timed, the overall ischemic time). For those recipients who are the beneficiaries of several prior palliative or corrective repairs, often the most expeditious approach—should the reoperative surgical field prove excessively hostile—is (1) performing the cardiectomy under deep hypothermic circulatory arrest simply to obtain a clearer sense of the underlying anatomy, (2) reconstruction to allow for bicaval (or tricaval) cannulation, and (3) reinstitution of bypass with rewarming to moderate hypothermia before the arrival of the donor heart.

This article addresses the 4 most common congenital abnormalities that require reconstruction at the time of transplantation, including: (1) management of the left superior vena cava, (2) management of transposition of the great arteries, (3) pulmonary artery reconstruction following prior congenital procedures, and (4) comprehensive reconstruction after prior hybrid-type palliation.

Left Superior Vena Cava

When a bridging or innominate vein exists, the recipient left superior vena cava (L-SVC) usually may be ligated. When no such bridging vein exists, the systemic venous return from the L-SVC (which could be considerable depending on its size) must be redirected toward the donor right atrium. Depending on the course of the L-SVC drainage within the heart, several different techniques may be applied. However, often the easiest is to reconstruct the L-SVC drainage using donor innominate vein, either sewn end to end or end to side to the recipient L-SVC (this is required if the patient has undergone a prior Glenn connection) (Fig. 1). If the L-SVC is considerable in size (eg, heterotaxy with interrupted IVC drainage), the creation of an intracardiac baffle may be helpful (Fig. 2). If the L-SVC drains to the coronary sinus through a “roofed” connection, some find it more direct simply to preserve this drainage when making the left atrial cuff, and utilize the enlarged coronary sinus as part of the bicaval or biatrial anastomosis to simplify reconstruction (Fig. 3). Some have further suggested that a left-sided Glenn connection (in the setting of bilateral Glens) can be left intact at the time of transplant with little clinical consequence, and moreover that the creation of a left Glenn can be an option if reconstruction to the systemic atrium is not feasible.

Transposition of the Great Arteries

The most common situation involving malposed great vessels is either (1) in the setting of adult Mustard or Senning patients or (2) patients with L-TGA and heart failure. Here, because of the more anterior-posterior

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orientation of the great vessels compared with the more left-right orientation of the donor arteries, it is helpful to adjust for the “twist” that results from using additional donor aorta or pulmonary artery (Fig. 4). Alternatively, adjusting the location of the recipient pulmonary arteriotomy can help to adjust for this discrepancy.

**Pulmonary Artery Reconstruction**

The most common anatomical distortion encountered is pulmonary artery stenosis from prior repairs. The most pronounced variant of this occurs in the setting of transplantation in a patient who has undergone a prior Fontan procedure (Fig. 5). Although isolated pulmonary artery stenosis is often best repaired with a simple patch, for those patients in whom multiple pulmonary arteriotomies are required, often utilization of bilateral branch pulmonary arterial tissue affords the most optimal reconstruction, as well as highest likelihood for appropriate growth over time. Naturally, those with discontinuous pulmonary arteries require prosthetic interposition to establish continuity.

**After Hybrid Interventions**

Increasingly, patients with cardiomyopathy, complex single ventricle disease, and aortic arch hypoplasia are undergoing hybrid procedures (ductal stenting, bilateral PA bands +/- atrial septal stent) as a bridge to transplantation. Reconstruction following stent and band removal can be challenging, and depending on issues of ischemic time, may often best be addressed before donor heart implant (Fig. 6).
Operative Technique

Figure 1  Systemic venous drainage from the donor left superior vena cava (L-SVC) without continuation to the coronary sinus (eg, after bilateral Glenn connection) can be brought back to the recipient right atrium most easily by utilizing additional donor innominate vein (IV) included at the time of procurement. In this rendering, the anastomosis of recipient L-SVC to donor IV is end to end; it can also be made end to side, in which case, often it is easiest to reduce distortion of the IV by using a vascular stapler to occlude the end of the IV with a staple line (rather than oversew). Note that this reconstruction either places the innominate vein behind (as represented) the aorta or in front. If the innominate vein lies behind the aorta, it is important to leave additional ascending aorta in the recipient and procure more from the donor to prevent compression of the IV by the ascending aorta. In contrast, if the IV lies anterior to the aorta, it is important to use little donor and recipient ascending aorta to shorten the ascending aortic anastomosis and thereby reduce the chance of stretching the overlying IV reconstruction. RSVC = right superior vena cava; Ao = aorta.
Figure 2  (A) If the connection of the recipient L-SVC is to the roof of the left atrium (and does not follow a “roofed” pathway to the coronary sinus [CS]), the systemic venous return from the L-SVC may be reconstructed as in Figure 1. However, if the donor innominate vein is not sufficient in length, or if the return from the L-SVC is going to exceed the capacity of the donor innominate vein (eg, heterotaxy and interrupted inferior vena cava with hemiazygos continuation), then other baffles can be created to redirect this systemic venous return.
Figure 2 (Continued) (B) The baffle may travel along the roof of the left atrium, above the pulmonary veins to a surgically created atrial septal defect.
Figure 2 (Continued) (C) This baffle may also be placed inferiorly to join the right atrium (RA) at the coronary sinus.
Figure 3  (A) If the native L-SVC drains via a “roofed” pathway to the coronary sinus (CS), it is simplest to keep this native connection intact. In this diagram, the course of the L-SVC is noted in dashed lines, as is the right atriotomy incision line (to the “bare area” on the right atrium) that is used for a biatrial anastomosis. R SVC = right superior vena cava; AO = aorta; PA = pulmonary artery.
Figure 3 (Continued) (B) When performing the recipient cardiectomy in this situation, it is important not to disrupt the L-SVC to CS pathway, and to trim the recipient atriotomy very close to the atroventricular valves as shown. The atria are then prepared for a simple biatrial anastomosis.
Alternatively, if a bicaval connection is preferred, the large CS can be left in situ connected to the recipient inferior vena cava (IVC) to make a large IVC cuff. Again, once the cardiectomy is completed, the connection is a simple standard bicaval anastomosis.
Figure 4  (A) Transposed great arteries may pose little problem for reconstruction. However, often their more anterior-posterior orientation (compared with a more rightward-leftward orientation of the donor heart) can result in a twist. Allowing for extra donor and recipient aorta and pulmonary artery can afford this twist without creating areas of stenosis.
Figure 4 (Continued) (B) However, should the donor or recipient tissue prove inadequate, it is often easier to simply transpose the location of the recipient pulmonary arteriotomy leftward. As shown, this aligns the great vessel connections to more easily match those of the donor heart.
One of the most common problems encountered is iatrogenic distortion of the branch pulmonary arteries from prior palliative repairs. After removal of the right, and sometimes left, superior vena cava following Glenn cavopulmonary connection, and additionally the Fontan connection along the inferior aspect of the PA, several deficiencies in pulmonary arterial tissue are evident. For the sake of reconstruction, often it is easier to enlarge the arteriotomy to encompass all of these smaller defects. RSVC = right superior vena cava; LSVC = left superior vena cava.
Figure 5 (Continued) (B) The donor pulmonary artery (ideally with some amount of bilateral branch pulmonary arterial tissue) can then be opened along its cranial aspect [inset], its edges beveled, and then used as an entire onlay patch. The advantage of this approach, in contrast to one in which each individual pulmonary arteriotomy is patched, is that the donor branch pulmonary arterial tissue serves not only to patch the defect but also to augment the overall lumen with “native” tissue and not prosthetic. In general, in this setting, the easiest order of anastomoses is caval or atrial, then pulmonary artery, and then aorta. PA = pulmonary artery; Ao = aorta.
Figure 6 (A) Patients who have undergone prior hybrid procedures, either as a bridge to decision or transplant, or as a primary therapy for hypoplastic left heart syndrome and its variants, present new challenges for reconstruction at the time of transplant. The most common scenario is depicted in (A) where bilateral pulmonary artery bands are placed as is a ductal stent (with or without a stent in the atrial septum).
At the time of transplantation, removal of the pulmonary artery bands often does not incur permanent distortion of the branch pulmonary arteries, and therefore the pulmonary artery anastomosis can proceed as usual. Removal of the ductal stent with adjacent descending aorta at its insertion requires reconstruction of the entire arch beyond the ductal insertion; for the purposes of future growth and geometric lie of the transverse arch, this often is best accomplished with extended donor aortic tissue.
Alternatively, if removal of the pulmonary artery bands creates distortion, a strategy as described previously for onlay patching of the branch pulmonary arteries is generally effective. In addition, if overall ischemic time concerns dictate, it can be more efficient to reconstruct the aortic arch and ductal insertion site with prosthetic tissue (eg, homograft) under significant hypothermia, then rewarm before arrival of the donor heart and proceed with the aortic anastomosis to the reconstructed ascending aorta as usual at mild hypothermia. In both examples, the transplant is shown as a biatrial connection.
Conclusions

Cardiac transplantation for end-stage congenital heart disease accounts for an increasing proportion of heart transplants performed worldwide. In particular, adults with congenital heart disease, most of whom have had several prior palliative procedures, provide new challenges requiring the expertise of both adult (acquired) and congenital heart surgeons. Recent reports suggest improving outcomes with transplantation for complex congenital heart disease, including those with “failed Fontan” physiology.8-10

For even the most complex reconstructions, often an efficient surgical strategy involves reconstructing the recipient anatomy so that it will allow for a standard biatrial or bicaval connection (rather than using adjunctive donor tissue to adjust for distortion). In this way, by performing most of the “reconstruction” during the donor organ transportation time, the total ischemic time (and in particular warm ischemic time) can be reduced substantially. Inevitably, anatomical variants and iatrogenic distortion may ultimately result in intraoperative anatomy that may not be in alignment with preoperative imaging and planning. Thus, having a systematic approach toward the congenital cardiac transplant reconstruction—as well as a robust armamentarium of alternative techniques to complete the repair—is a key feature for overall success.

References