

# 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 anesthesia-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.<sup>1,2</sup>

For congenital cardiac transplant candidates, a standard, systematic approach toward surgical planning is recommended.<sup>3</sup> 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) reinstatement 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 Glenns) 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).

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