Surgical technique for heart transplantation: a strategy for congenital heart disease

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Summary

The standard techniques for orthotopic heart transplantation often require certain adjustments when the procedure is carried out for complex congenital heart disease. This is because of both the unusual anatomy and possible distortions caused by previous surgery. Such technical adjustments have been described in various published reports over the years. Those reports, when combined, do cover the full spectrum of the technical difficulties that may be encountered, whether the defects are in their original form or altered by surgery, such that no cardiac malformation or distortion would prohibit transplantation. However, those reports are comprehensive only when combined. None of the individual reports addresses all the possible technical challenges. Consequently, the available information is somewhat fragmented. In addition, the generic aspect of the described technical strategies is not always given the emphasis that it deserves. Indeed, occasionally a technique may be presented as a specific solution for a specific malformation, without necessarily pointing out that the same technique may be applied to other hearts with different overall pathologies but which share that specific malformation. The aim of this review article was to combine all the available published information in one article in a manner that collectively points out that the same technique may be applied to other hearts with different overall pathologies but which share that specific malformation. This leaves little room for improvement. Nevertheless, the existing literature has two weaknesses. First, those reports are comprehensive only when combined. None of the individual reports addresses all possible morphological challenges. Consequently, the available information is somewhat fragmented. Second, the generic aspect of the described techniques is not always given due emphasis: often a specific technique is presented as a specific solution for a specific malformation, without necessarily pointing out that the same technique may be applied to other hearts with different overall pathologies, but which share that specific malformation. For example, it would be useful to point out that the transplantation technique for hypoplastic left heart syndrome is also applicable whenever there are aortic arch anomalies independently of hypoplastic left heart syndrome.

INTRODUCTION

Two surgical techniques for orthotopic heart transplantation are widely practised. One is the classic technique described by Lower and Shumway consisting of four anastomoses, namely the left and right atrial, pulmonary arterial and aortic anastomoses [1]. The other is a modification of the classic technique whereby the recipient’s right atrium is completely removed and the donor’s heart is implanted using bicaval anastomoses [2]. The latter has become very popular and many surgeons use it exclusively. However, both techniques require certain adjustments in 60–75% of patients requiring transplantation for congenital heart disease because of unusual anatomy and/or possible distortions caused by previous surgery [3, 4].

The necessary technical adjustments have been described in various reports [3, 4–30]. These reports, when combined, do address the full spectrum of possible morphological challenges such that no cardiac malformation or distortion due to previous surgery would prohibit heart transplantation. The only exceptions are severe hypoplasia of pulmonary arteries or veins, when heart–lung transplantation may be indicated [14, 30]. This leaves little room for improvement.

Nevertheless, the existing literature has two weaknesses. First, those reports are comprehensive only when combined. None of the individual reports addresses all possible morphological challenges. Consequently, the available information is somewhat fragmented. Second, the generic aspect of the described techniques is not always given due emphasis: often a specific technique is presented as a specific solution for a specific malformation, without necessarily pointing out that the same technique may be applied to other hearts with different overall pathologies, but which share that specific malformation. For example, it would be useful to point out that the transplantation technique for hypoplastic left heart syndrome is also applicable whenever there are aortic arch anomalies independently of hypoplastic left heart syndrome.
OBJECTIVE

We aim to combine all published technical adjustments in one article in a manner that constructs a simple but comprehensive and generic system of decision-making that may be applied to any heart in order to determine the exact transplantation technique needed. This is possible for two reasons:

(i) Only a few anatomical sites are technically relevant, namely the points of anastomosis between the donor’s heart and the recipient (the points of systemic and pulmonary venous returns, and the two great vessels). The rest of the intracardiac morphology does not affect the operation and may be ignored. Even dextrocardia is not of great importance as it permits the standard transplantation technique [5, 6, 8]. However, with dextrocardia, the pericardium should be widely opened into the left pleura, and its right-hand edge pulled up and attached to the anterior chest wall, in order to minimize the tendency of the transplanted heart to assume a right-sided position [6].

(ii) Each of these anatomical sites can present difficulties in only a few ways, and each of those few difficulties has a well-described solution already.

Therefore, the exact technical adjustments required in each case may be worked out by sequential assessment of the anastomotic sites alone.

This approach facilitates the planning of the operation from the purely morphological point of view. However, the technical challenges of transplantation for congenital heart disease extend beyond morphology. Those non-morphological technical challenges, although well known, do deserve a brief mention prior to the description of our proposed sequential assessment.

NON-MORPHOLOGICAL TECHNICAL CHALLENGES

There are four main non-morphological challenges, namely those of resternotomy, dissection of adhesions, the possible need for reconstruction of the anastomotic sites and the complications of cyanosis when applicable.

The sternotomy may be hazardous because patients with end-stage congenital heart disease often have both a distended right heart and extensive adhesions caused by previous surgery. This problem may be managed, on occasion, by establishing cardiopulmonary bypass via the peripheral vessels prior to the resternotomy. However, this approach may be difficult in patients with congenital heart disease since the peripheral vessels may be distorted or blocked due to multiple previous catheter studies, or (in children) too small for cardiopulmonary bypass. One option may be axillary cannulation if the size of the patient and that of the axillary vessels allow this. But often the cannulation must be within the chest.

Adhesions from multiple previous operations are often extensive and dense. The dissection often takes a long time, with significant repercussions on the management of the donor. On occasion, the adhesions and distortions are so extensive that the excision of the recipient’s heart may need to be carried out under deep hypothermic circulatory arrest [3, 8].

Reconstruction of one or more of the anastomotic sites may be required, after the excision of the recipient’s heart, for satisfactory implantation of the donor’s heart. This is best done using living tissue (recipient’s or donor’s) rather than prosthetic material that may increase the risk of bleeding and infection. Avoiding prosthetic material may be facilitated by excising only what is absolutely necessary from the recipient, and requesting the harvesting team to harvest as much tissue as possible from the donor (extra length of superior caval and innominate veins, the great vessels and pericardium). Such communication with the harvesting team is essential. Nevertheless, occasionally, extensive recipient tissue excision is required, and the surgeon is forced to use foreign material for reconstruction.

Cyanotic patients usually have extensive collateral circulation, making the conduct of bypass difficult and increasing the risk of bleeding. The risk of bleeding is further increased by the coagulopathy that is often seen in cyanotic patients with high haematocrit.

THE PROPOSED SEQUENTIAL ASSESSMENT

We propose the following sequence:

(i) What is the pattern of pulmonary venous return?
(ii) What is the pattern of systemic venous return?
(iii) What is the atrial morphology (situs)?
(iv) What is the morphology of the main pulmonary artery and its two main branches?
(v) What is the morphology of the ascending aorta and aortic arch?

The pattern of pulmonary venous return

The possible anomalies are as follows:

(i) various forms of total or partial anomalous pulmonary venous return;
(ii) severe stenosis/hypoplasia or atresia of the pulmonary veins.

This is one of the two above-mentioned exceptions where isolated heart transplantation is not possible, and heart–lung transplantation may be indicated [14, 30].

With the exception of severe stenosis/hypoplasia/atresia of the pulmonary veins, the pulmonary venous drainage to the left atrium is almost never a problem for transplantation. When there is a left atrium, it is a midline structure receiving the pulmonary veins. If the pulmonary veins do not connect to the left atrium, they are immediately adjacent to it and can be easily connected to it surgically. Indeed, such correction is likely to have been carried out prior to the transplant procedure by anastomosing the venous collector that receives the anomalous veins to the back of the left atrium in a side-to-side fashion. In such cases, the transplant technique requires no adjustment. Uncorrected anomalous pulmonary venous drainage must be corrected at the time of heart transplantation, assuming that heart transplantation is not contraindicated due to pulmonary hypertension. The correction is done by the same technique except that the common pulmonary venous collector is anastomosed to the back of the left atrium of the donor’s heart, and the vertical vein that connects this pulmonary venous collector to the systemic veins is ligated (Fig. 1) [4, 30].

One form (the commonest) of partial anomalous pulmonary venous drainage deserves specific mention since the anomalous vein is not adjacent to the left atrium. This is when the right
superior pulmonary vein drains into the superior caval vein. This anomaly is also likely to have been corrected previously in one of two manners. When the opening of the anomalous vein is at the junction of the superior caval vein with the right atrium, a baffle is created (usually using fresh autologous pericardium) directing the anomalous pulmonary venous blood through the right atrium and atrial septum towards the left atrium. When transplanting such a heart, the only adjustment required is to make sure that the left atrial anastomosis maintains and does not distort the opening of the baffle into the left atrium. This is achieved by not resecting the native atrial septum (to which the baffle was sutured previously) at the time of extraction of the recipient's heart (Fig. 2). Apart from this precaution, the standard transplantation techniques (classic or bicaval) may be applied. Occasionally, however, the original correction may have been done by the Warden procedure. This is chosen when the opening of the anomalous pulmonary vein is higher up on the superior caval vein where a baffle would cause superior caval obstruction. In such cases, the entire opening of the superior caval vein is baffled through the right atrium and atrial septum towards the left atrium. Then, the superior caval vein is transected just above the level of the opening of the anomalous pulmonary vein and is anastomosed to the right atrial appendage. Such a case is managed similarly to the simple baffle: with minimal resection of the recipient's atrial septum so that the opening of the baffle into the left atrium is not distorted. However, the right heart must be managed by the bicaval technique.

If such a malformation has not been previously corrected, a baffle directing the anomalous vein to the left atrium via the interatrial communication can be constructed at the time of transplantation.

**The pattern of systemic venous return**

The possible anomalies are as follows:

(i) presence of additional left superior caval vein with a bridging (innominate) vein;
(ii) presence of additional left superior caval vein without a bridging vein;
(iii) interruption of the inferior caval vein, with azygos continuation connecting to the left superior caval vein;
(iv) absence of the right superior caval vein.

A left-sided superior caval vein is not a problem if there is a bridging (innominate) vein connecting it to the right-sided superior caval vein. The left superior caval vein can then be ligated and divided below the level of the bridging vein [4, 8]. In the absence of a bridging vein, however, some technical adjustments are needed. Several techniques have been described, such as preserving the coronary sinus when this structure drains the left superior caval vein, and various forms of re-routing (baffling) of the left superior caval return (when this does not drain into the coronary sinus) through the left atrium towards the right atrium (Fig. 3A) [7, 8]. However, the easiest technique is an extracardiac adjustment whereby the left superior caval vein of the recipient is anastomosed to the innominate vein of the donor while its distal stump (which is connected to the back of the left atrium or coronary sinus) is ligated or oversewn (Fig. 3B) [11]. If the donor's organ arrives without the innominate vein, a prosthetic conduit may be used to establish continuity between the right and left caval veins.

In the rare instances of interruption of the hepatic segment of the inferior caval vein with hemi-azygos continuation connecting to the left superior caval vein, an intracardiac adjustment is required in order to maintain the drainage of the inferior caval vein into the right atrium [11]. An extracardiac adjustment, as in the above instances of bilateral superior caval veins, would be inadequate since this would impose the drainage of both (superior and inferior) caval systems on the superior caval vein of the transplanted heart, which is unlikely to be sufficiently big for this. In addition, the left-sided superior caval vein in such instances is generally much bigger than usual, resulting in too much size discrepancy for a good anastomosis with the donor's innominate vein. Therefore, the intracardiac approach is advised. If the left superior caval vein drains into the coronary sinus, the coronary sinus must be preserved and incorporated into the new (transplanted) right atrial chamber. If the coronary sinus is unroofed, it must be repaired with a patch. Also, the site of its tributaries (draining the myocardium of the recipient's heart) must be oversewn. If the left superior caval vein opens independently into the left atrium, then a baffle must be constructed diverting it through the left atrium to the right atrium. This may be done using autologous or donor's pericardium, or native

![Figure 1: Anastomosing the pulmonary venous collector (PVC) to the back of the left atrium (LA).](image1)

![Figure 2: The anomalous right superior pulmonary vein is baffled (asterisk) through the right atrium (RA) and atrial septal defect, toward the left atrium (LA).](image2)
Atrial morphology (situs)

The possible variations are as follows:

(i) usual atrial arrangement (situs solitus), which presents no added difficulty;
(ii) ambiguous atrial arrangement (situs ambiguous: right and left atrial isomerism);
(iii) mirror-image atrial arrangement (situs inversus);
(iv) distortion of the atria due to previous surgery (Mustard, Senning and Fontan procedures).

With situs ambiguous, the patient usually has a single atrium that receives all systemic and pulmonary veins. This may be managed by using a patch to partition the atrium in a manner that directs the systemic venous blood towards the right and the pulmonary venous blood towards the left [3–5, 7]. However, if the required partition is simple (in one plane), there is no need for a patch: the right side of the donor’s left atrial wall is minimally trimmed, and this excess tissue can be sewn to the posterior wall of the recipient’s common atrium, thus creating an interatrial septum [4, 8].

In situs inversus, the recipient’s heart is excised, leaving a right-sided morphologically left atrial cuff and a left-sided morphologically right atrial cuff in the recipient. It is important to remove as little of the left-sided morphologically right atrial tissue as possible. These transposed atrial cuffs carry with them the openings of their respective veins (i.e. the pulmonary and systemic venous drainage systems are also transposed). Various techniques have been described in order to address this, such as a modification of the Senning procedure and a translocation technique for the morphologically right atrium [3–6, 30]. However, the easiest technique is an extracardiac baffle (Fig. 5): The recipient’s superior caval vein (which is left sided) is disconnected from the left-sided morphologically right atrium. This morphologically right atrial tissue is completely separated from the right-sided morphologically left atrial cuff by dissecting into the interatrial groove and cutting its free wall off the left atrium. The interatrial septum remains attached to the right-sided morphologically left atrial cuff. This results in a very generous muscular flap attached to the opening of the left-sided inferior caval vein. This flap is incised on its medial aspect down to, and on the medial aspect of, the left-sided inferior caval vein down to the level of the diaphragm. This manoeuvre requires a period of circulatory arrest so that the left-sided inferior caval venous cannula may be temporarily removed. This flap is folded towards the right, across the midline, onto the pericardium to which it is sutured, thus creating a tunnel directing the left-sided inferior caval venous blood rightward to the site corresponding to that of a normal right-sided inferior caval vein.

The recipient’s morphologically left atrial cuff is anastomosed to the back of the donor’s left atrium in the usual fashion. The systemic venous return is established using the bicaval anastomotic technique with the following adjustments. The recipient’s inferior caval baffle is anastomosed to the inferior caval entry point of the donor’s right atrium. The recipient’s superior caval vein is anastomosed to the donor’s innominate vein in an end-to-end fashion (Fig. 3B). Alternatively, the donor’s superior

Figure 3: Various techniques of dealing with the presence of an additional left-sided superior caval vein. (A) intra-atrial re-routing (baffling), (B) the easier extracardiac adjustment. RSVC: right-sided superior caval vein; L SVC: left-sided superior caval vein; IV: innominate vein; RA: right atrium.

Figure 4: A method of dealing with absence of the right superior caval vein. SVC: superior caval vein; LSVC: left-sided superior caval vein; IV: innominate vein; RA: right atrium.
caval vein is anastomosed to the recipient’s innominate vein in an end-to-side fashion (Fig. 4).

The inferior caval venous baffle sits easily below the transplanted heart for two reasons [4, 6]. First, the size disparity between the recipient’s heart (usually large because of end-stage heart failure) and the donor’s heart means that the pericardial sac of the recipient can accommodate more than just the transplanted heart. Second, this venous conduit sits in the natural indentation of the atrioventricular groove.

Distortion of both atria is encountered in patients who have had a previous atrial repair for transposition of the great arteries (Senning or Mustard procedures). Distortion of the right atrium is also seen in all patients who have had a Fontan procedure. In cases of previous Senning or Mustard procedures, almost the entire recipient’s heart is excised, including the atrial baffle, leaving behind only a cuff of left atrial wall containing the openings of the four pulmonary veins. This cuff is anastomosed to a matching defect created on the back of the donor’s left atrium. The caval veins are managed by the bicaval anastomosis method.

A previous Fontan operation distorts the right atrium regardless of the exact form of Fontan that was performed. In addition, if the Fontan were of the extracardiac total cavopulmonary connection type, then both caval veins would have already been disconnected from the right atrium and diverted towards the pulmonary arteries. The left atrium is usually normal. In such cases, the obvious solution for heart transplantation is the bicaval anastomosis technique.

(iii) severe hypoplasia or absence of extrapulmonary branches of the pulmonary arteries. This requires heart-lung transplantation, as mentioned previously.

The key to the management of these challenges is the fact that the pulmonary artery, in both donor and recipient (if present), will be in the midline at some point in its path. Therefore, the donor’s and recipient’s pulmonary arteries may be easily adjusted to each other as long as sufficient pulmonary artery (beyond the bifurcation) has been harvested from the donor [3, 10].

If the main pulmonary artery is absent or distorted by previous surgery, direct anastomosis of the individual branch pulmonary arteries may be carried out (Fig. 6A). However, distortions from previous surgery are often localized, and a simple localized patch repair may be sufficient (Fig. 6B).

Morphology of the ascending aorta and aortic arch

Challenges may arise from:

(i) aortic hypoplasia/interruption/coarctation, although these are likely to have been repaired previously;
(ii) distortion from previous surgery, such as arch repair, Damus-Kaye-Stansel procedure (the pulmonary trunk anastomosed to the ascending aorta), repair of truncus arteriosus, and the arterial switch operation with or without the Lecompte manoeuvre;
(iii) spatial relationship between the aorta and the pulmonary artery may be abnormal. Here, the possibilities are transposition of the great arteries and various forms of double-outlet ventricle. In these conditions, the two great arteries may occupy a wide range of different positions with respect to each other;
(iv) right aortic arch.

The management of the ascending aorta is facilitated by the fact that the aorta is located to the right of, and anterior to, the
pulmonary artery at the point of exit from the pericardium, regardless of both where it arises from and how it is related to the pulmonary artery within the pericardium. There are only three exceptions to this rule: previous surgery involving deliberate reconstruction of the right ventricular outflow tract to the right of the aorta, a previous Lecompte manoeuvre (transferring the pulmonary arterial bifurcation to the front of the ascending aorta, as done typically in the arterial switch operation) and situs inversus. Therefore, apart from these three exceptions, all ascending aortic anomalies can be managed with adequate mobilization of the recipient’s aorta and use of extra length of donor’s aorta [3–5, 10].

Even with situs inversus, or previous reconstruction of the right ventricular outflow tract to the right of the aorta, the aortic anastomosis may still be managed similarly with adequate mobilization of the recipient’s aorta and use of extra length of donor’s aorta. However, it may be necessary to shift the pulmonary arterial anastomosis slightly sideways to the left in order to avoid a kink caused by the aorta [30]. This is achieved by adjusting the pulmonary anastomotic site along the confluence of the right and left pulmonary arteries (Fig. 7). However, if there is no such confluence, or it has been distorted by previous surgery, then direct anastomoses between the donor’s and recipient’s branch pulmonary arteries are needed (Fig. 6A). In such cases, the sideways shift in the position of the main pulmonary artery can be achieved by adjusting the lengths of the two branch pulmonary arteries. If a Lecompte manoeuvre had been done previously, this should simply be reversed after the excision of the recipient’s heart [3].

Proximal aortic arch anomalies should be treated by the so-called hemi-arch repair: oblique anastomosis between the donor’s ascending aorta and the undersurface of the recipient’s aortic arch (Fig. 8) [30]. Distal aortic arch pathology (interruption or coarctation), should be treated separately from the transplant in the course of the procedure (i.e. repair the arch, then do the transplant). As with all surgery on the aortic arch, periods of circulatory arrest would be needed. Therefore, the patient should be cooled accordingly.

HEART-LUNG TRANSPLANTATION

For heart-lung transplantation, the sequential analysis is even easier. The surgeon is only interested in a tracheal anastomosis, an aortic anastomosis and the systemic venous anastomoses. These are the only sites that require assessment. Atrial anomalies and anomalies of pulmonary arteries and veins are irrelevant as these structures are excised.

OUR OWN EXPERIENCE

We retrospectively studied all paediatric and adult congenital heart transplant cases in our database. There were a total of 95 patients from 1993 till 2011. All procedures had been performed by surgeons specialized in congenital heart disease. The median age was 2 years (range 2 days to 19 years). Seventy percent of them had undergone cardiac surgery previously. Congenital
malformations were the principal diagnosis in 49 cases (52%). One-year survival was 77% for the whole group (70% for the congenital patients, 85% for the others).

Among the 49 congenital cases, 46 (94%) required at least one technical adjustment to the standard transplantation technique—a percentage that is noticeable superior to that presented in the literature (60–75%). However, the distribution of the patients in terms of which segment of the heart and great vessels were affected was not homogeneous. There were significantly more arterial anomalies when compared with venous malformations, and there was only one with mirror-image atrial arrangement (situs inversus). Indeed, in 45 of the 49 cases (92%), the affected segments were the great arteries: hypoplastic left heart syndrome, transposition of the great arteries, congenitally correct transposition and common arterial trunk (truncus arteriosus).

In each case, we used our model to predict the necessary technical adjustments, and then compared this with what was actually carried out. The concordance was 100%—in every case, the predicted technique coincided exactly with the one that was used.

**COMMENTS**

In this review, we offer a generic and comprehensive strategy that helps determine how the standard heart transplantation techniques may be adjusted when the procedure is being carried out for congenital heart disease. It is based entirely on existing techniques. We offer no new technique, but a new way of viewing and cataloguing them.

The proposed strategy, when tested retrospectively on our own series, proved to be exact (100% concordance between the predicted and the actual techniques). This supports its utility as an accurate guide. Such a guide might be particularly useful for surgeons who are not experts in congenital heart disease. One may argue that such transplants should be carried out only by surgeons specialized in congenital heart disease. Indeed, this has largely been the case so far. However, the expected increase in the number of such patients [31–33] is unlikely to allow this.

This model also underscores the vital importance of close communication between the team operating on the recipient and the harvesting team, because many of the technical problems require the harvesting of extra donor tissue.

**Conflict of interest:** none declared.

**REFERENCES**


