Operative Techniques in Thoracic and Cardiovascular Surgery

# Extracardiac Conduit Fontan Procedure

Scott M. Bradley, MD

Staged palliation culminating in a successful Fontan procedure is the current surgical goal for most patients with a functional single ventricle. Since its introduction in 1968, the Fontan procedure has undergone many modifications. The extracardiac conduit Fontan procedure (ECC) was introduced in the late 1980s.[1,2] The ECC includes direct connection of the superior vena cava (or cavae) to the pulmonary arteries, and placement of a tube graft outside of the heart, from the inferior vena cava to the pulmonary arteries.

The ECC has several advantages (both theoretical and real) over other modifications of the Fontan procedure, such as the intra-atrial lateral tunnel (LT). The ECC limits atrial suture lines to the superior and inferior vena caval transection sites, and the fenestration site (if one is used). Following an ECC, no portion of the atrium is exposed to high (pulmonary artery) pressure, thus avoiding atrial wall stretch and distention. These characteristics have the theoretical advantage of decreasing postoperative atrial arrhythmias. When performed after a previous bidirectional superior cavopulmonary shunt, the ECC avoids incisions and suturing in the region of the sino-atrial node. This has the theoretical advantage of decreasing sinus node dysfunction. The ECC avoids potentially thrombogenic prosthetic material in the pulmonary venous atrium, as well as potential baffle leaks through intra-atrial suture lines. The ECC also leaves the coronary sinus to drain to the low pressure (pulmonary venous) atrium. Addition or modification of a fenestration can be performed without cardiopulmonary bypass, either in the operating room or in the intensive care unit.

The ECC is very versatile and can be performed in essentially all anatomic situations. This includes patients in whom other approaches might be difficult, such as those with heterotaxy syndrome and anomalies of systemic and pulmonary venous drainage, or apicocaval juxtaposition. In the event of subsequent heart transplantation, the ECC is fairly easily taken down to accommodate a bicaval implantation of the donor heart. This is in contrast to patients who have had a previous hemi-Fontan procedure and LT Fontan, who require more extensive pulmonary artery reconstruction at the time of transplantation.

Division of Cardiothoracic Surgery, Medical University of South Carolina, Charleston, South Carolina.

Address reprint requests to Scott M. Bradley, MD, Division of Cardiothoracic Surgery, Medical University of South Carolina, 96 Jonathan Lucas St., Charleston, South Carolina 29425. E-mail: bradlesm@musc.edu.

The ECC can be undertaken with a variety of support strategies. These range from performance without cardiopulmonary bypass[3-5] to the use of deep hypothermic circulatory arrest.[6,7] The following illustrations show use of cardiopulmonary bypass, without aortic cross-clamping. This approach allows a technically optimal and expeditious operation. The entire central pulmonary arteries, including the previous superior cavopulmonary anastomosis, are well exposed, without encumbering clamps. The ability to use cardiotomy suckers provides a measure of safety. Cardiopulmonary bypass times are generally short, while hypothermia and myocardial ischemia are avoided. Intracardiac procedures are performed at the time of the superior cavopulmonary anastomosis. In patients presenting for ECC who still require a procedure such as atrioventricular valve or aortic arch repair, a staged approach is considered: the operation requiring aortic cross-clamping is performed first; the ECC is performed at a later date, as an isolated procedure without myocardial ischemia.

A variety of conduits have been used in the ECC Fontan. These have included Dacron and Gore-Tex (W.L. Gore & Associates, Flagstaff, AZ) tube grafts, aortic homografts, partial or complete tubes of pedicled autologous pericardium, and tissue-engineered grafts. The following illustrations show the use of a Gore-Tex tube graft. Gore-Tex has the advantages of ready availability in a variety of sizes, and adequate flexibility. It would likely be relatively easy to redissect in the event of replacement, although this has not yet been necessary in our experience. Like other prosthetic grafts, Gore-Tex lacks growth potential. This limitation is typically approached by operating once the patient is large enough (roughly 15 kg) to accept a graft adequate for an adult's inferior caval flow (20- to 22-mm-diameter).

As illustrated in the following figures, the ECC is typically performed following a previous superior cavopulmonary anastomosis. Routine preoperative work-up includes echocardiography and cardiac catheterization to define ventricular function, atrioventricular and semilunar valve function, systemic outflow tract obstruction, and pulmonary artery anatomy, pressure, and resistance. As a routine, aortopulmonary collaterals are neither identified nor coil occluded at the preoperative catheterization.[8] In the occasional patient with marginal hemodynamics, coil occlusion may be helpful when performed either shortly before or early after the Fontan procedure. Preoperative CT angiography is increasingly used to define arterial and venous anatomy, and the proximity of these structures to the sternum.


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# Operative Technique

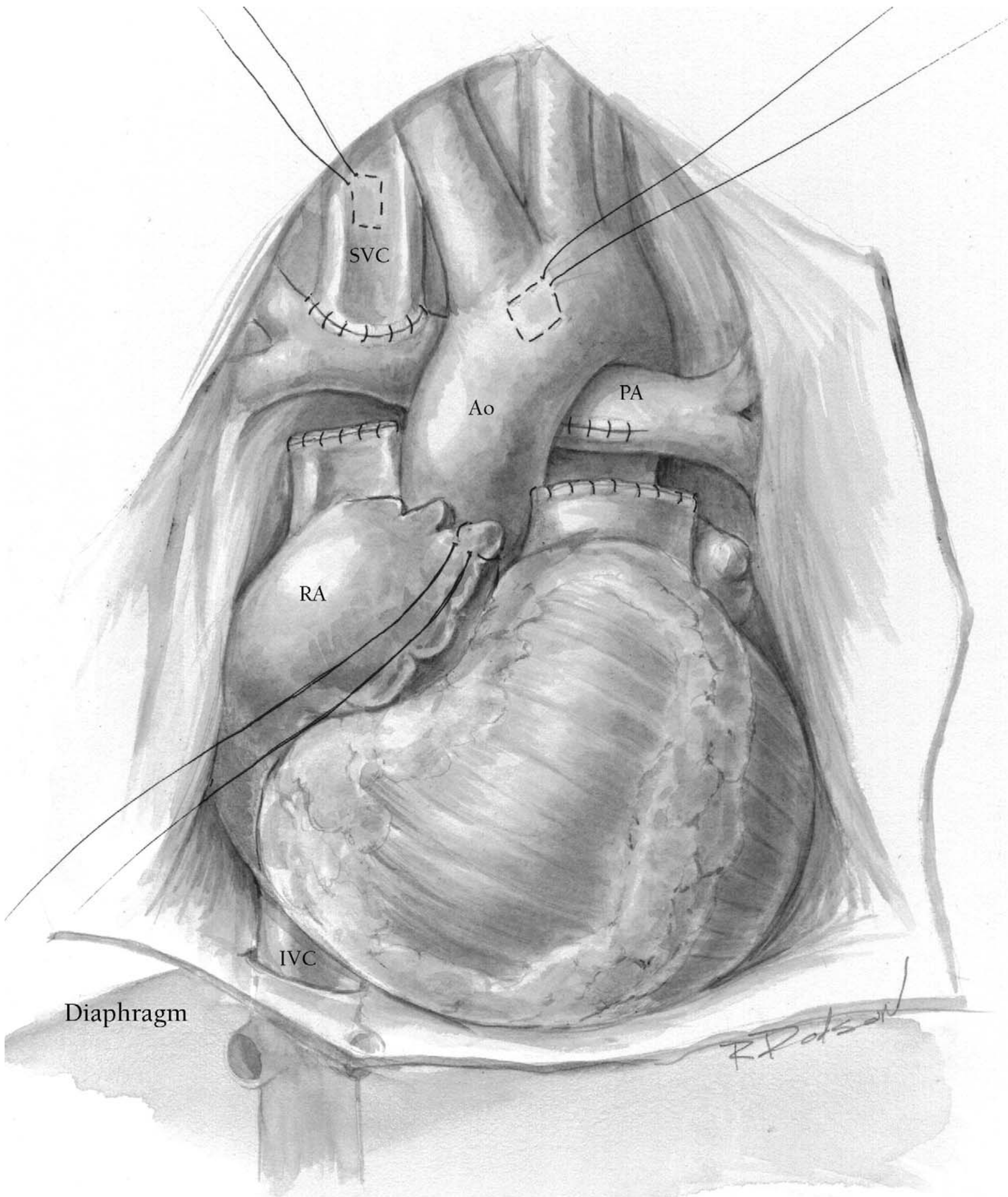



**Figure 1** The illustrations depict a patient who has had a previous bidirectional superior cavopulmonary anastomosis. Exposure is via a repeat median sternotomy. The pleural spaces are opened and adhesions are taken down to permit placement of Blake drains in both pleural spaces. Dissection includes enough of the ascending aorta and superior vena cava to cannulate, the two divisions of the right pulmonary artery, the central and left pulmonary artery behind the aorta, the right atrium, and the inferior vena cava. To avoid injury to the right phrenic nerve, cautery is avoided and sharp dissection with scissors is used along the rightward aspect of the superior vena cava and around the divisions of the right pulmonary artery. Purse-string sutures are shown in the aorta, the superior vena cava, and the area of the right atrial appendage. Aprotinin is used routinely, but not initiated until enough of the cardiac structures have been exposed to permit cannulation in the event of an anaphylactic reaction. Ao = aorta; IVC = inferior vena cava; PA = pulmonary artery; RA = right atrium; SVC = superior vena cava.

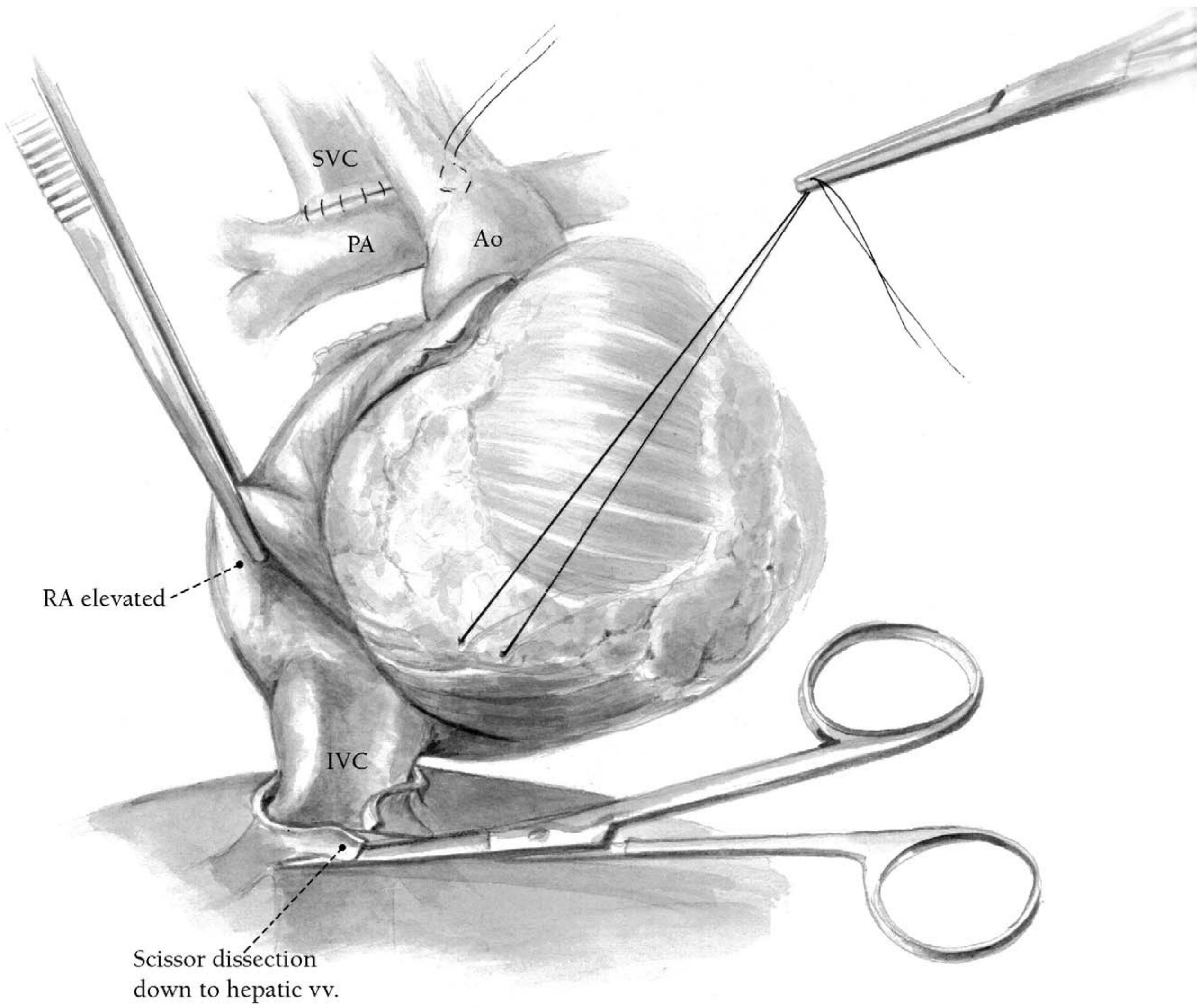



**Figure 2** Exposure of the inferior vena cava is facilitated by freeing up the diaphragmatic surface of the ventricle and placing a retraction suture in the acute margin of the heart, avoiding coronary arteries. The inferior vena cava is dissected into its diaphragmatic hiatus, until the hepatic veins are visualized. Placement of a malleable retractor on the diaphragm (not shown) may be helpful in the dissection of the inferior vena cava. Ao = aorta; IVC = inferior vena cava; PA = pulmonary artery; RA = right atrium; SVC = superior vena cava; vv = veins.

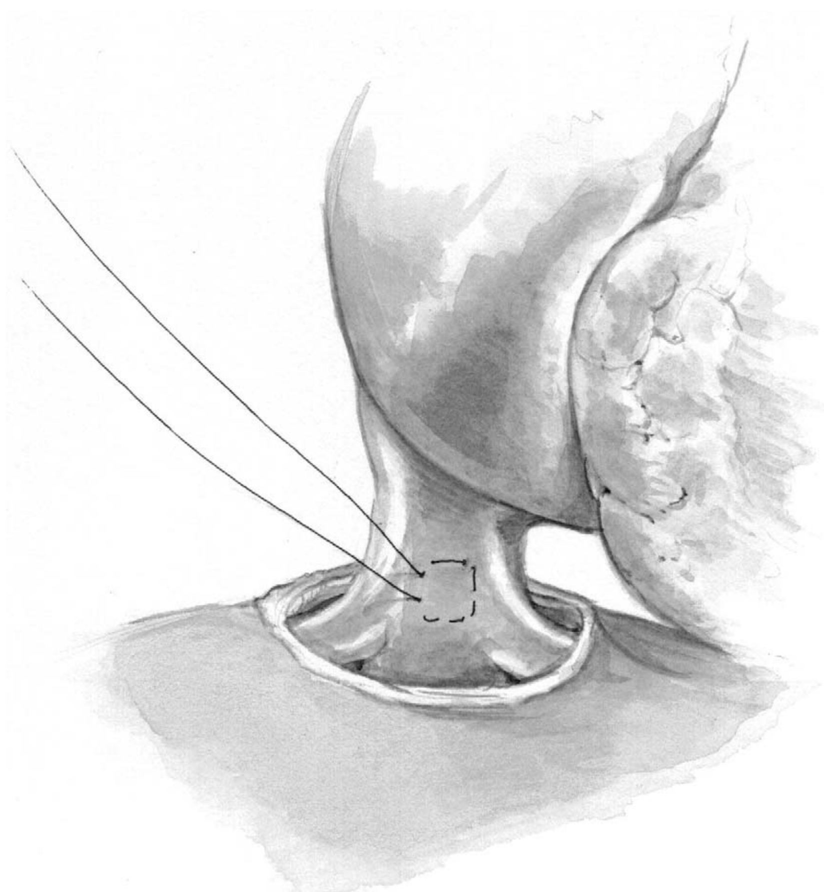


**Figure 3** A purse-string suture in the inferior vena cava is placed inferiorly, at the entry of the hepatic veins. Cannulation of the inferior cava via the femoral vein is an alternative in patients weighing more than approximately 20 kg.

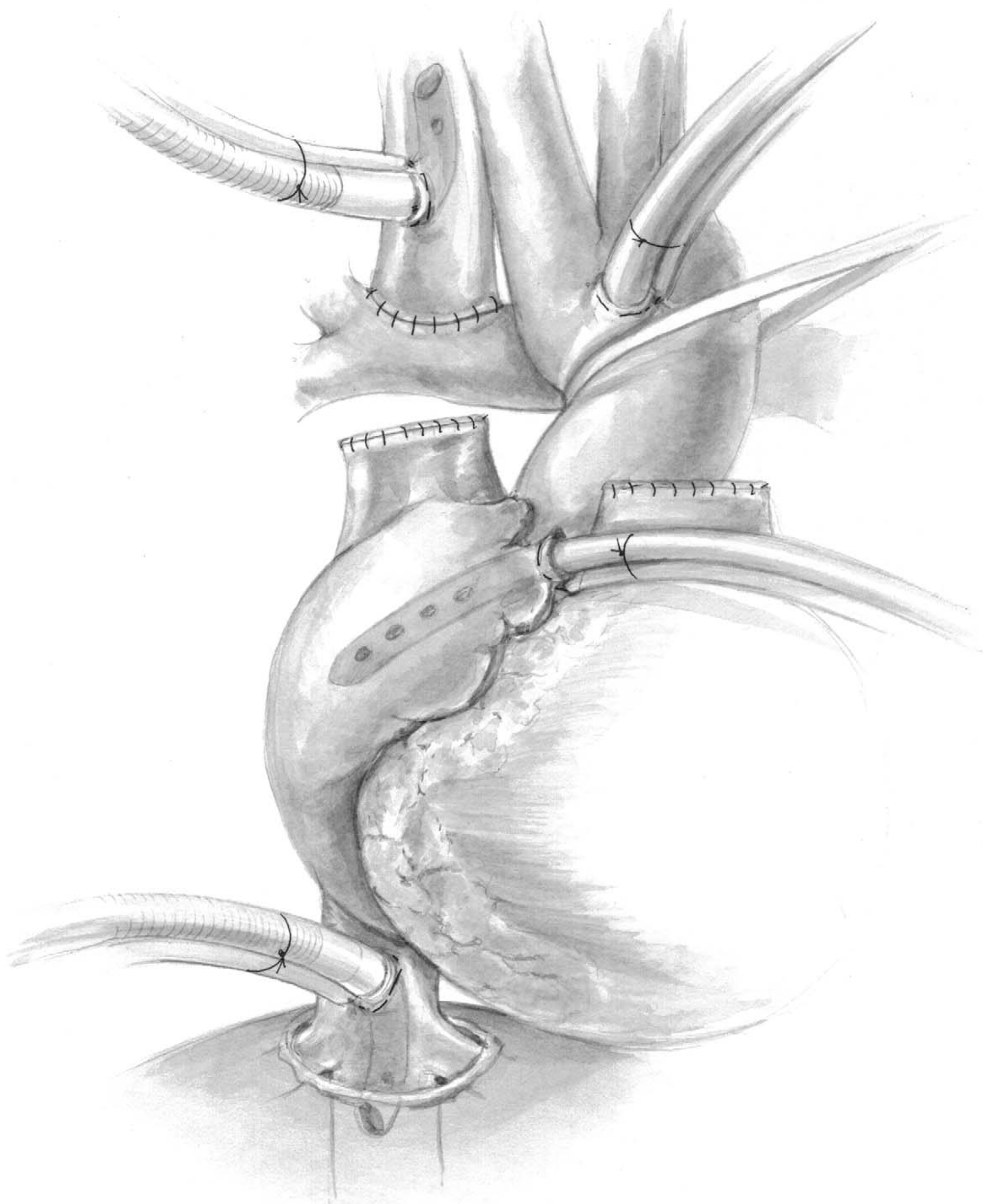


**Figure 4** Following heparinization, the aorta is cannulated, and right-angled cannulae (DLP, Medtronic, Minneapolis, MN) are placed in the venae cavae. Although not always used, a sump placed in the atrium can be useful to drain pulmonary venous return, which can be copious, depending on the extent of aortopulmonary collaterals. Decompression of the atrium can further help exposure of the inferior vena cava for its subsequent division (below). Exposure of the central and left pulmonary arteries is facilitated by circumferential dissection of the ascending aorta, and placement of an umbilical tape for retraction, as shown. If the patient has had a previous Norwood procedure with homograft reconstruction of the neoaorta, the pulmonary arteries are generally quite adherent to the posterior aspect of the neoaorta. This dissection can be completed following initiation of bypass. Dissection in this area is facilitated by leaving a small piece of Preclude Gore-Tex pericardial membrane between the neoaorta and pulmonary arteries at the time of the superior cavopulmonary anastomosis. The patient's temperature is maintained above 32°C, and the heart is maintained beating throughout the procedure.

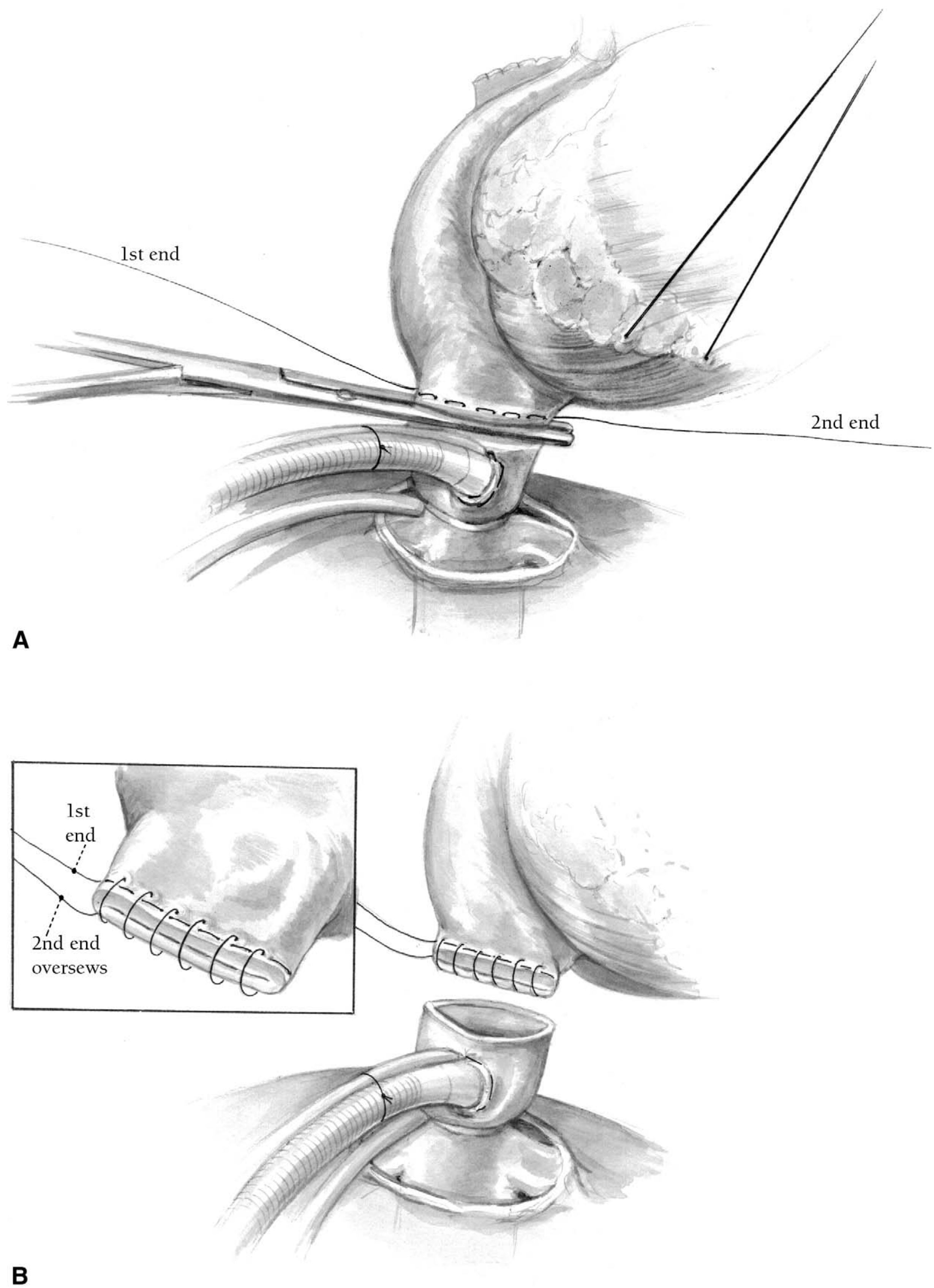



**Figure 5** (A) The previously placed retraction stitch and the atrial appendage sump purse-string sutures are pulled superiorly and leftward, to expose the inferior vena cava. The cava is circumferentially dissected up to the heart and the right lower pulmonary vein. A caval cannula snare is placed. A straight vascular clamp is placed across the cava, leaving enough tissue inferiorly to sew to the conduit. One end of a 4-0 prolene suture on a BB needle is sewn as a running horizontal mattress across the atrial side of the clamp. It is important to place this suture line in atrial tissue, avoiding the coronary sinus and right atrioventricular groove; accurate identification of atrial tissue is aided by observing the sequential beating of atrium and ventricle before applying the clamp. (B) The inferior vena cava is divided by cutting with a knife on the inferior side of the clamp. The clamp is then removed and the second end of the prolene suture is sewn over and over the atrial side of the transected cava. Care is again taken medially to avoid passing this suture into the area of the right atrioventricular groove.

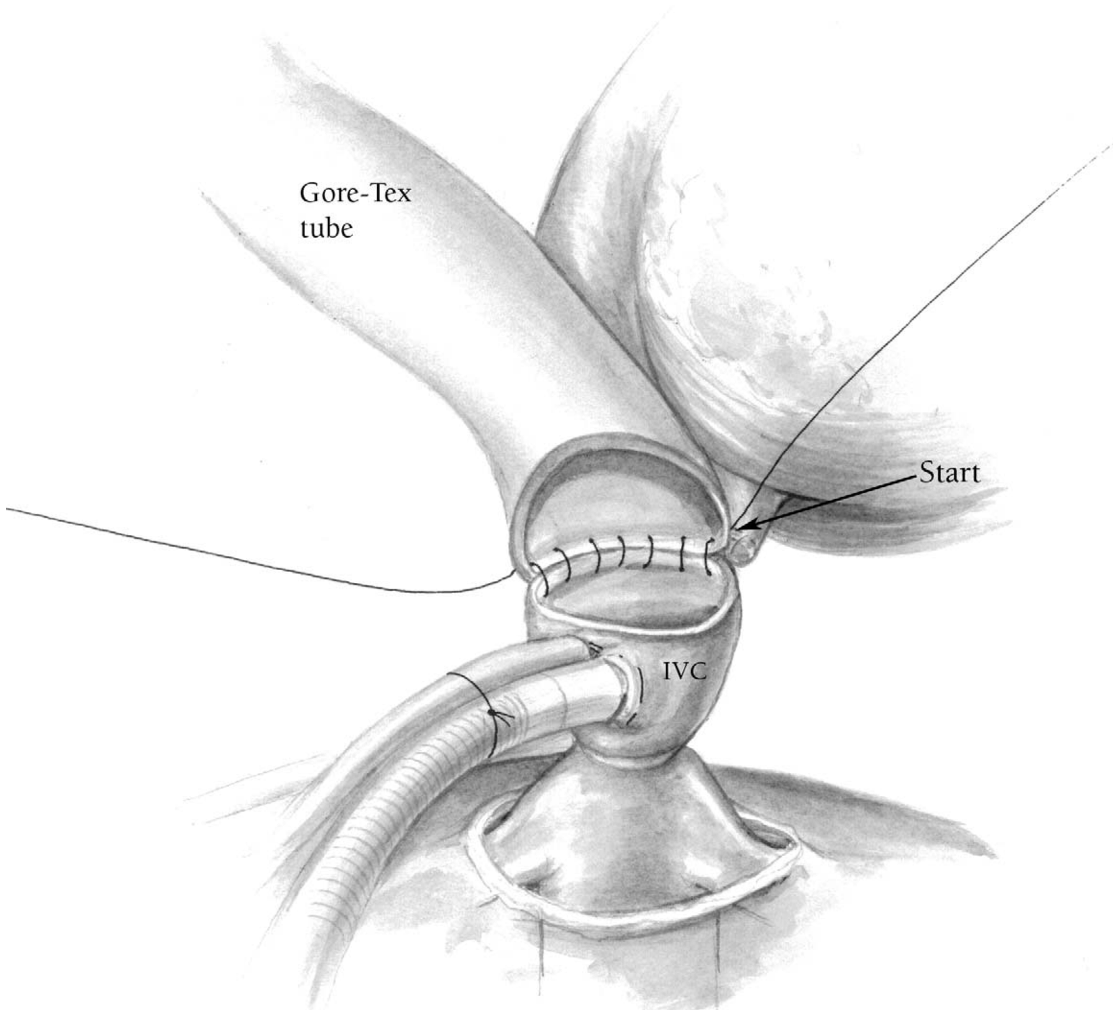
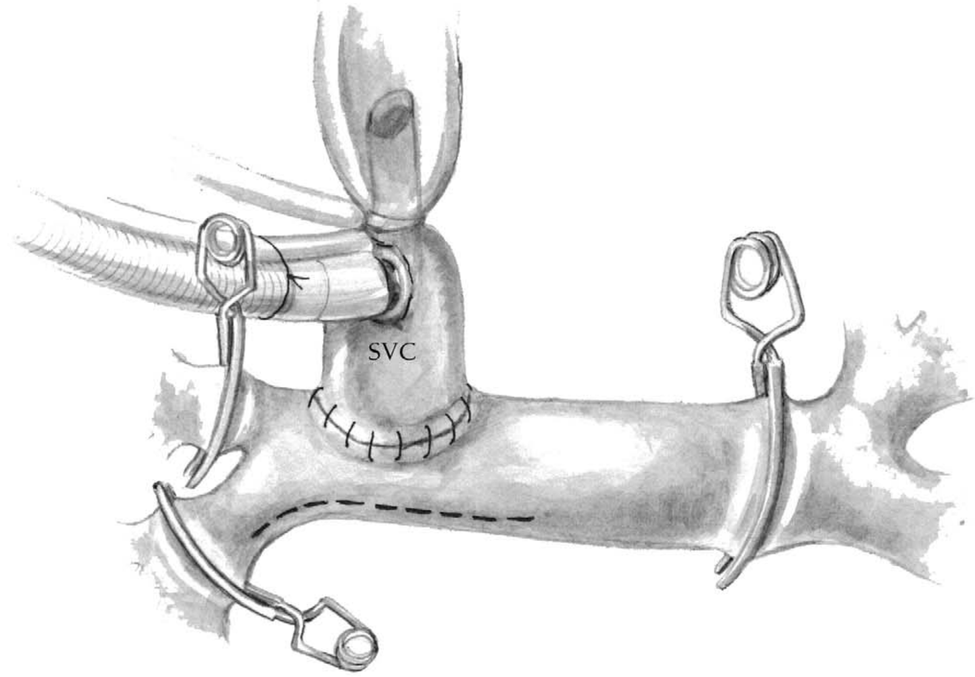



**Figure 6** The caval cannula snare is placed on traction inferiorly to elevate the transected cava into the field. Prominent Eustachian valve remnants are resected, and the external circumference of the cava is clearly identified. A Gore-Tex tube graft, generally 20 to 22 mm in diameter, is cut straight across, and sewn end-to-end to the transected inferior vena cava. A 4-0 or 5-0 Gore-Tex suture is used, sewing across the back wall from the inside, and the front wall from the outside. At the completion of this suture line, traction is released on the caval snare, the appendage sump snare, and the acute margin retraction suture, allowing the heart and inferior vena cava to fall back to their natural positions in the mediastinum. IVC = inferior vena cava.

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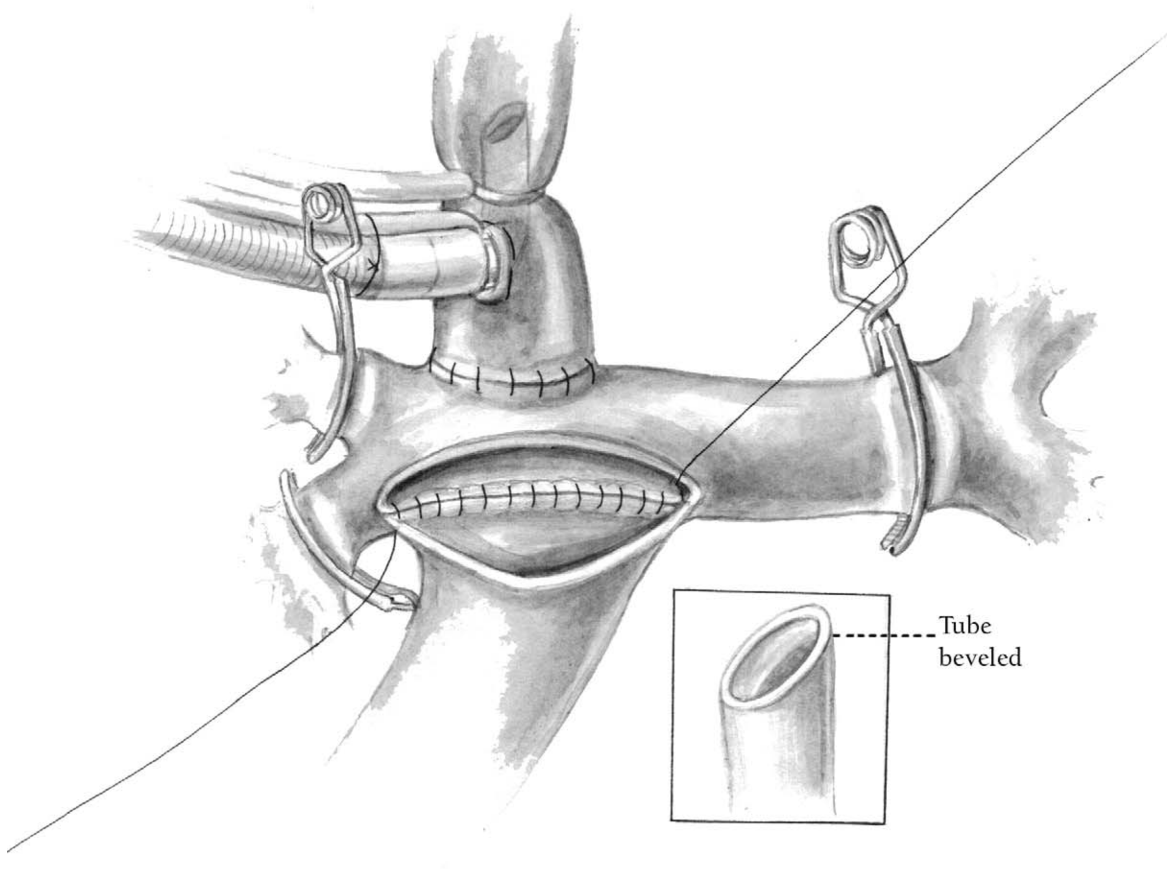



B

**Figure 7** (A) With the superior vena cava snared, neurosurgical clips (Codman & Shurtleff, Raynham, MA) are placed on the left pulmonary artery and the divisions of the right pulmonary artery. An incision is made on the inferior aspect of the pulmonary arteries, extending from the lower division on the right toward the left pulmonary artery. This incision generally lies opposite the superior cavopulmonary anastomosis. If the left pulmonary artery is difficult to expose and control, return from the left is managed with a flexible pump sucker introduced once the pulmonary arteries are opened. SVC = superior vena cava. (B) The length of the conduit is carefully assessed from the inferior caval anastomosis to the lower division of the right pulmonary artery. The conduit is cut on a slight bevel and sewn to the opening in the pulmonary arteries. A 6-0 Gore-Tex suture is used, sewing across the back wall from the inside, and the front wall from the outside. SVC = superior vena cava.

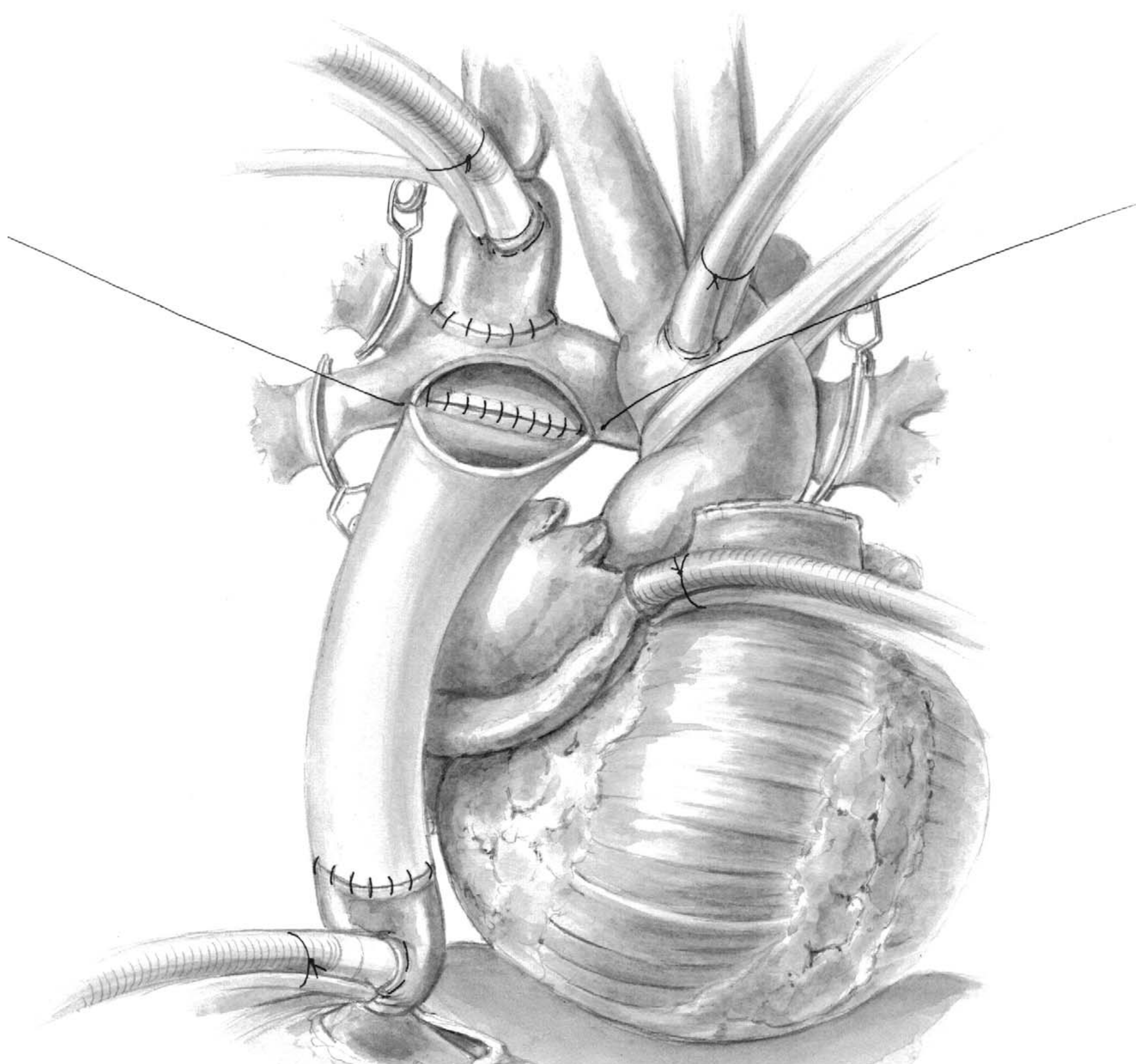
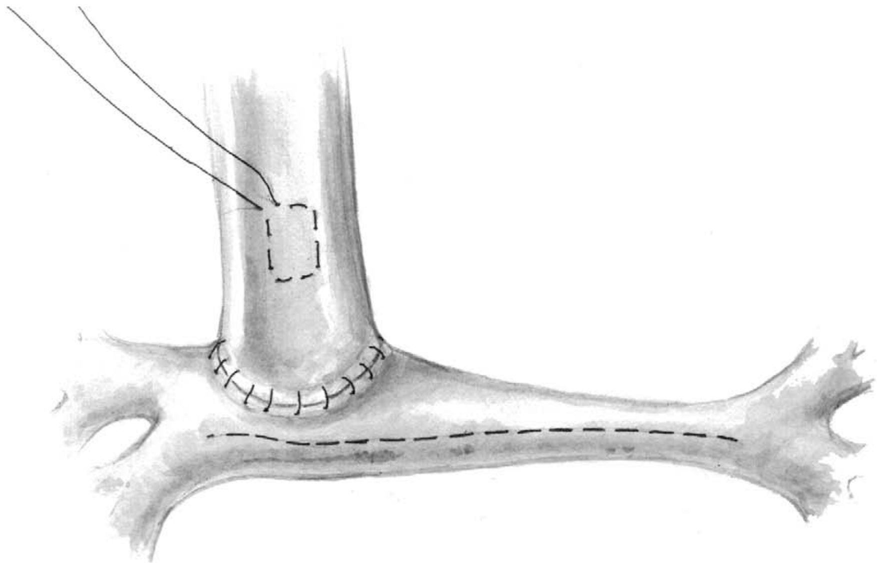


**Figure 8** The conduit should lie in a slight curve, without distortion of the right pulmonary artery, or compression of the right upper pulmonary vein, which lies immediately posterior.



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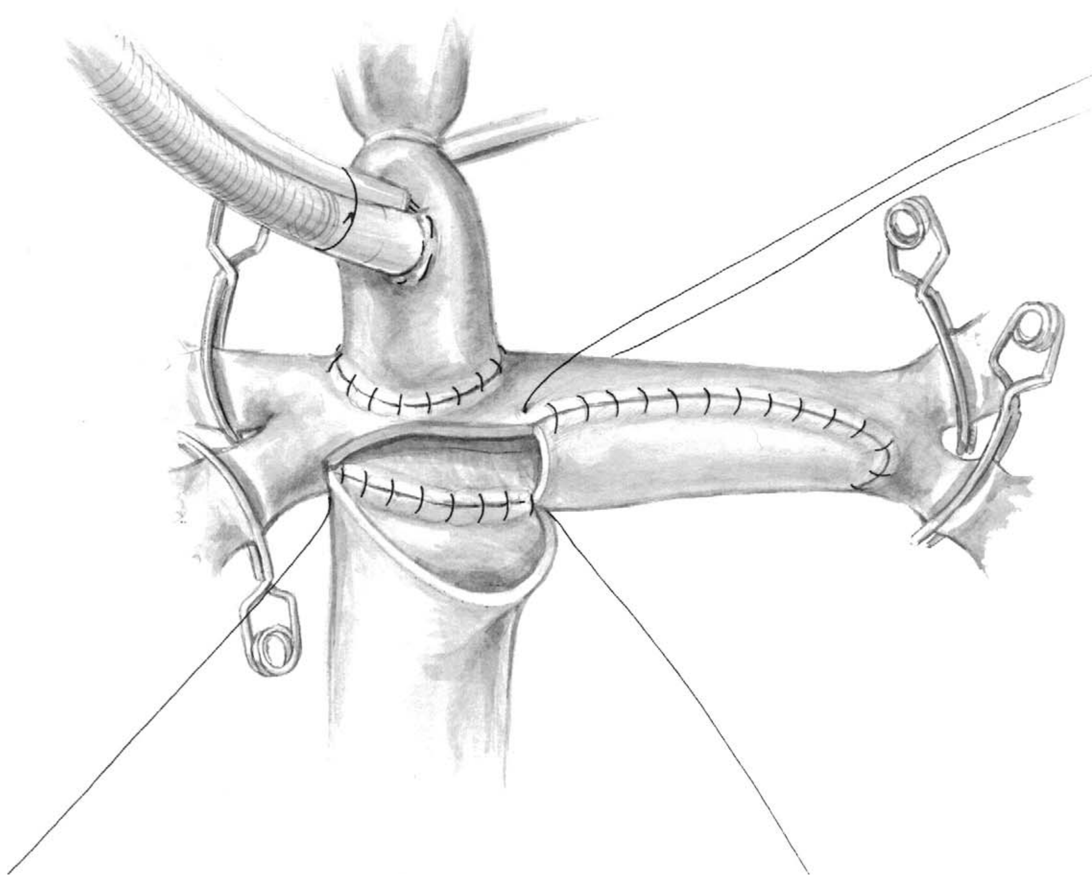

B

**Figure 9** (A) In the event of significant stenosis or hypoplasia of the left pulmonary artery, the artery is completely exposed to its branch points behind the aorta. The pulmonary artery incision will now extend from the lower division on the right, as far to the left as necessary to relieve all stenoses. (B) The conduit can be directly sewn to the length of the pulmonary artery incision by cutting the conduit in a long bevel (not shown). However, the reconstruction may have a better lie if a separate, bullet-shaped patch of bovine pericardium or Gore-Tex is initially sewn into the leftward aspect of the incision. This leaves an elliptical opening opposite the superior vena cava, to which the conduit is sewn after cutting it with essentially no bevel.

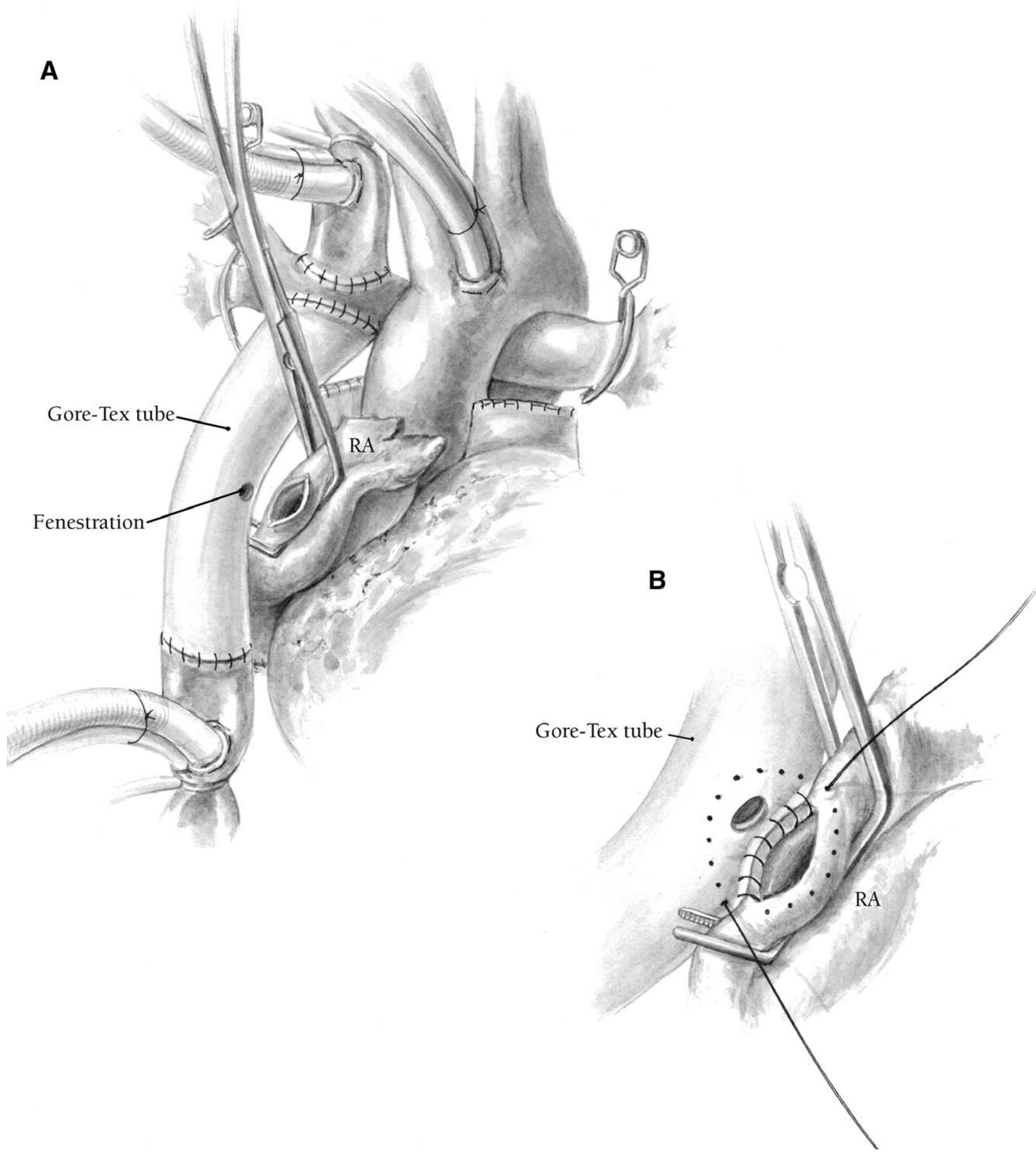


**Figure 10** A fenestration can be placed as a routine (our preference), or selectively, once hemodynamics are assessed after weaning from cardiopulmonary bypass. One effective technique is illustrated: a hole is placed in the side of the conduit using a punch. The size of the hole is generally 4 mm if the patient weighs <15 kg, and 5 mm if >15 kg. A side-biting clamp is placed on the lateral wall of the right atrium, staying anterior and inferior to the location of the sinoatrial node. The electrocardiogram is observed to ensure maintenance of P-waves. An incision is made in the atrium and sewn side-to-side to the conduit, taking partial thickness bites of the Gore-Tex, away from the edge of the punch hole. A closing snare is left around the fenestration (see below). RA = right atrium.

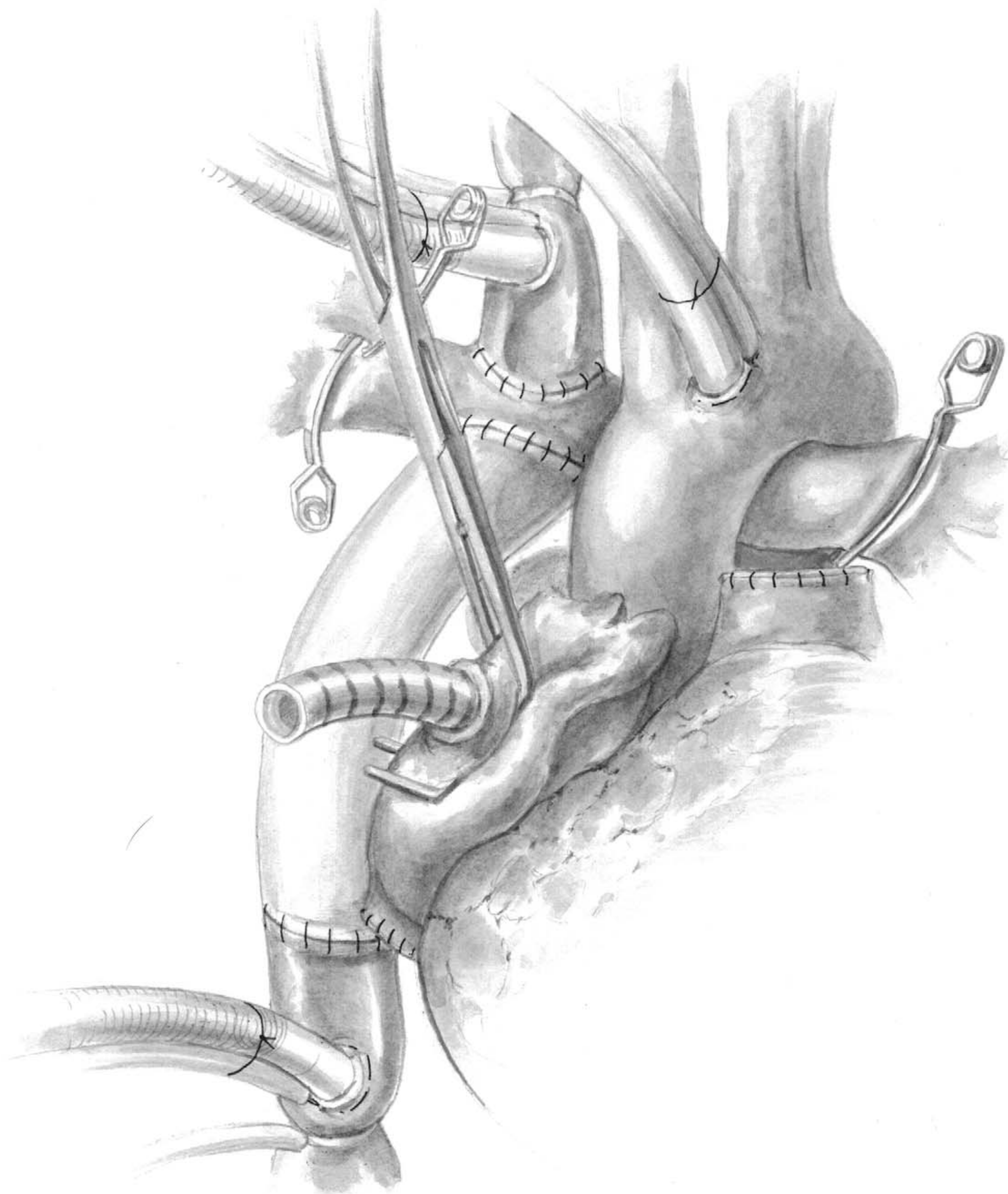

**Figure 11** An alternative technique of fenestration places a ringed Gore-Tex tube graft between the conduit and atrium. The diameter of the fenestration graft is generally 5 mm if the patient weighs $<15$ kg, and 6 mm if $>15$ kg. The atrial end is sewn using a side-biting clamp. One of the graft's rings is incorporated in the anastomosis, so as to stent open the connection to the atrium.

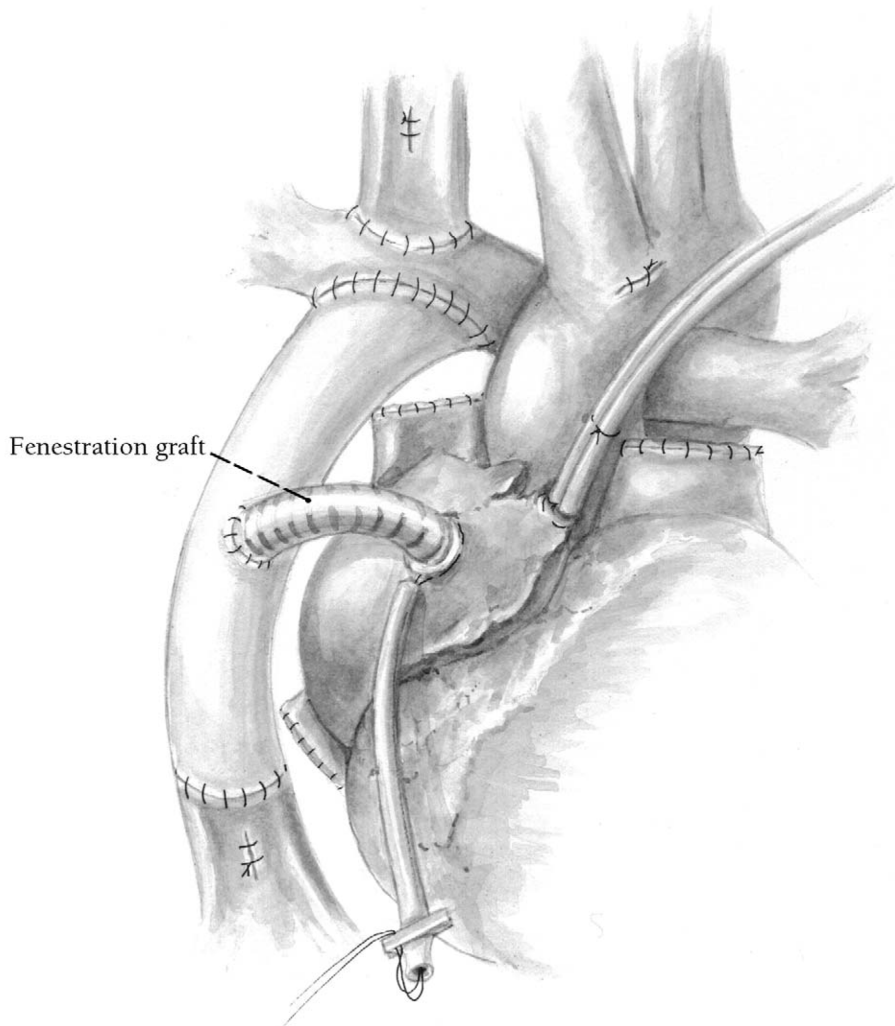


**Figure 12** The fenestration graft is generally 2 to 3 cm in length and lies in a curve from the Fontan conduit to the atrium. A closing snare (2-0 prolene) is placed around the atrial end of the fenestration graft and tacked to the atrial wall with fine sutures. The snare suture is placed in a snugger (a stiff, no. 10 red rubber catheter), clipped to the snugger, and buried under linea alba at the inferior end of the sternotomy incision. Should it be desired to close the fenestration, this is performed in the catheterization laboratory 9 to 12 months following surgery. The fenestration is test-occluded with a balloon catheter. If hemodynamics remain suitable, the end of the closing snare is retrieved through a 1-cm incision using local anesthesia. The snare is pulled tight, clipped to the snugger, and reburied under linea alba. This technique allows fenestration closure without placing an additional foreign body in the bloodstream.

At the conclusion of the procedure, a transthoracic monitoring line is left in the common atrium via the appendage sump site. Temporary atrial and ventricular pacing wires are also placed.

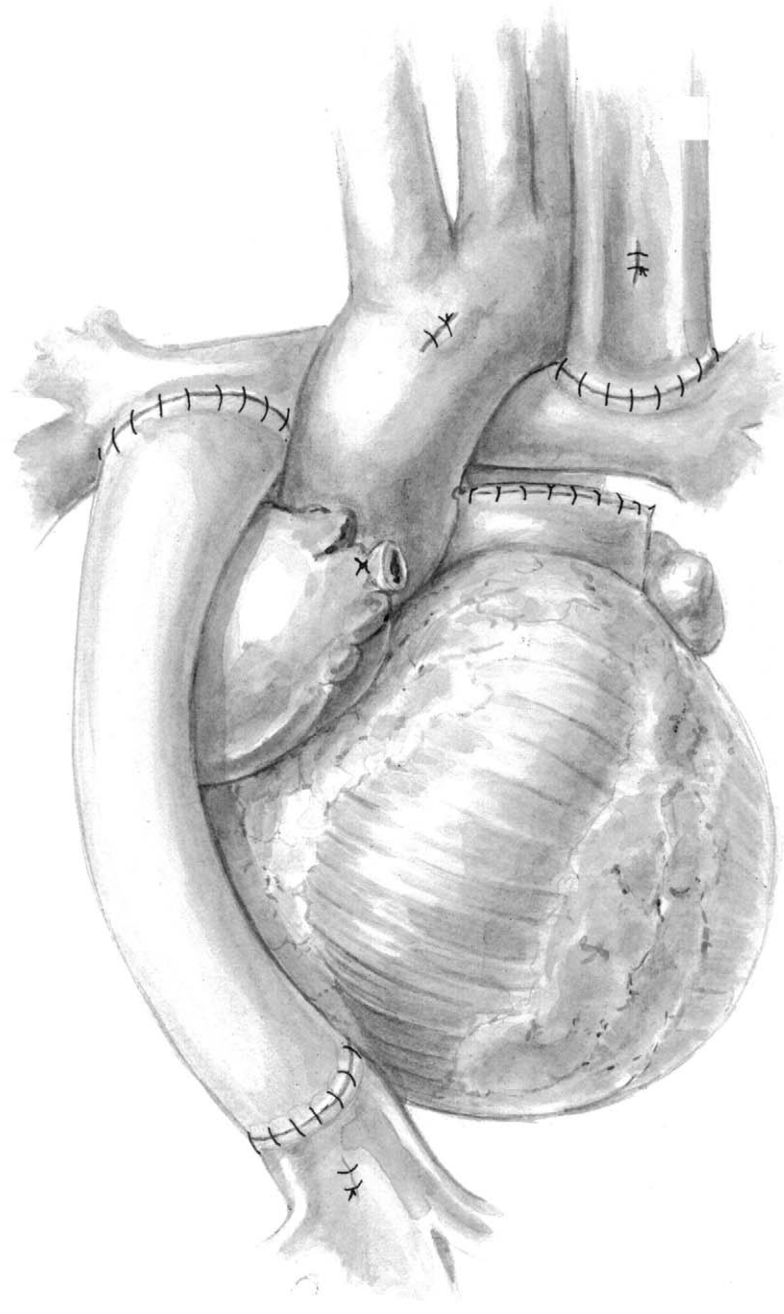

**Figure 13** In the setting of apicocaval juxtaposition, it can be difficult to fashion an intra-atrial baffle from the inferior vena cava to the pulmonary arteries without interfering with either the atrioventricular valve or the pulmonary veins. An ECC can be placed in a smooth curve from the mobilized inferior vena cava to the pulmonary artery opposite the apex of the heart (the right pulmonary artery in this illustration).


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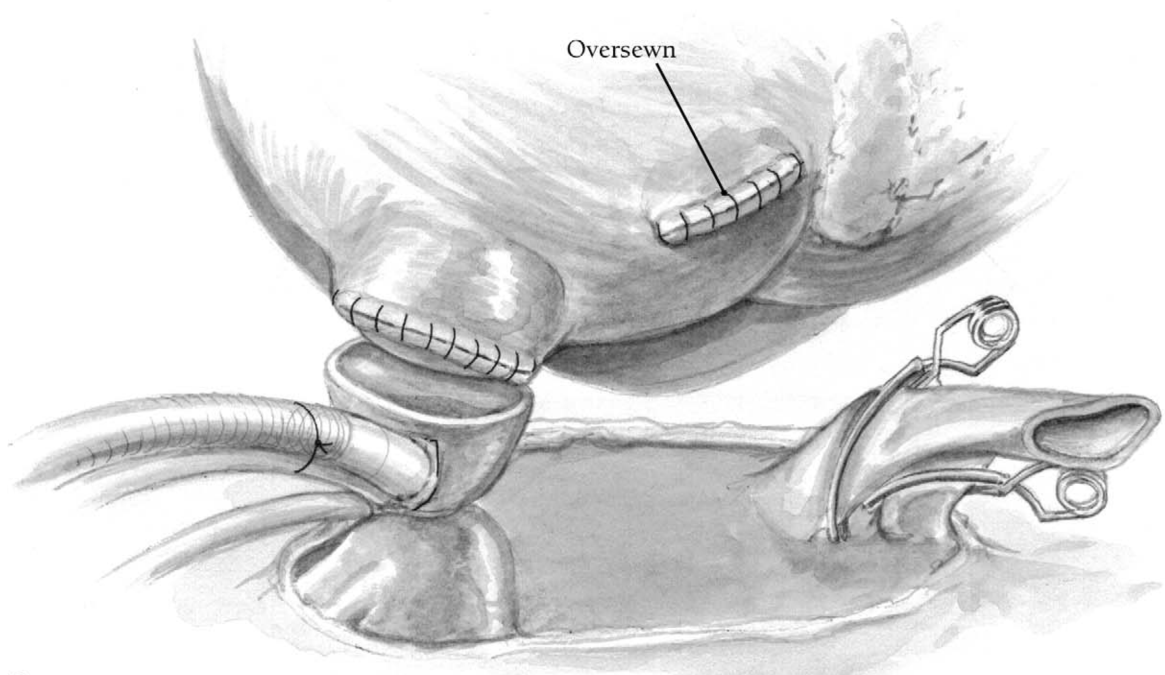
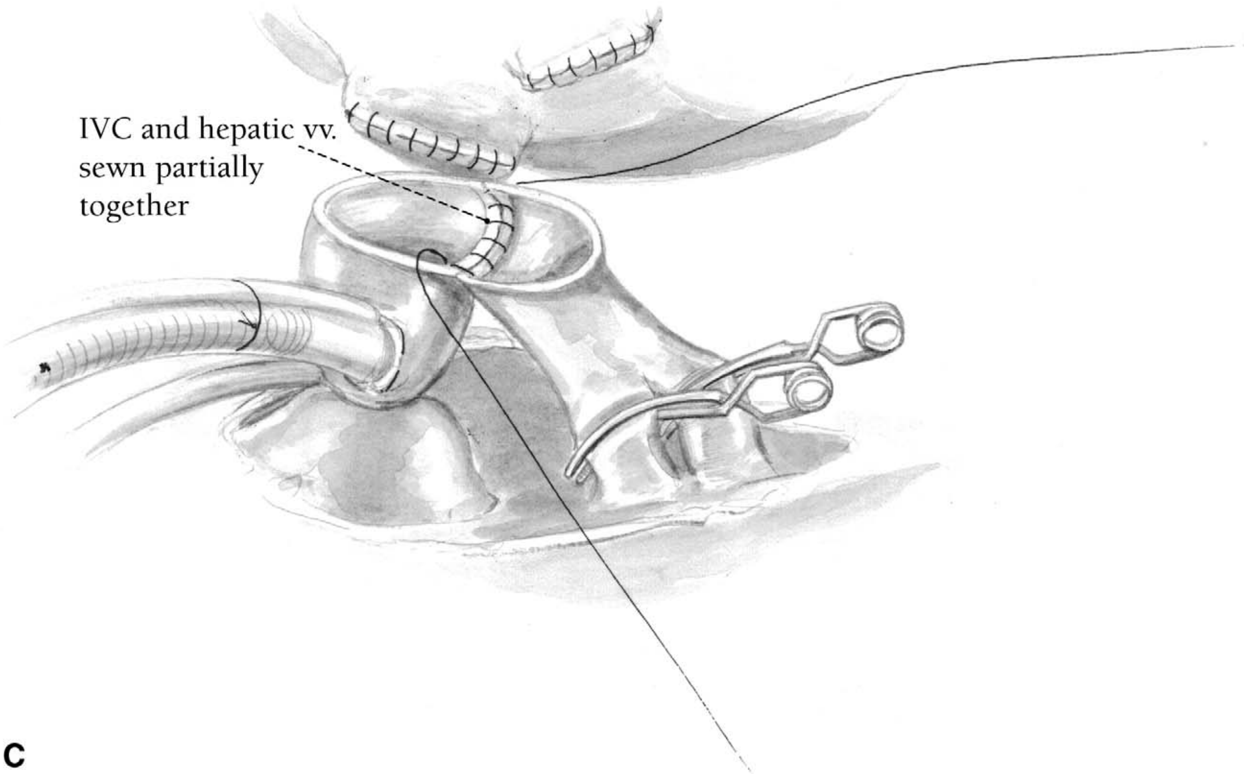
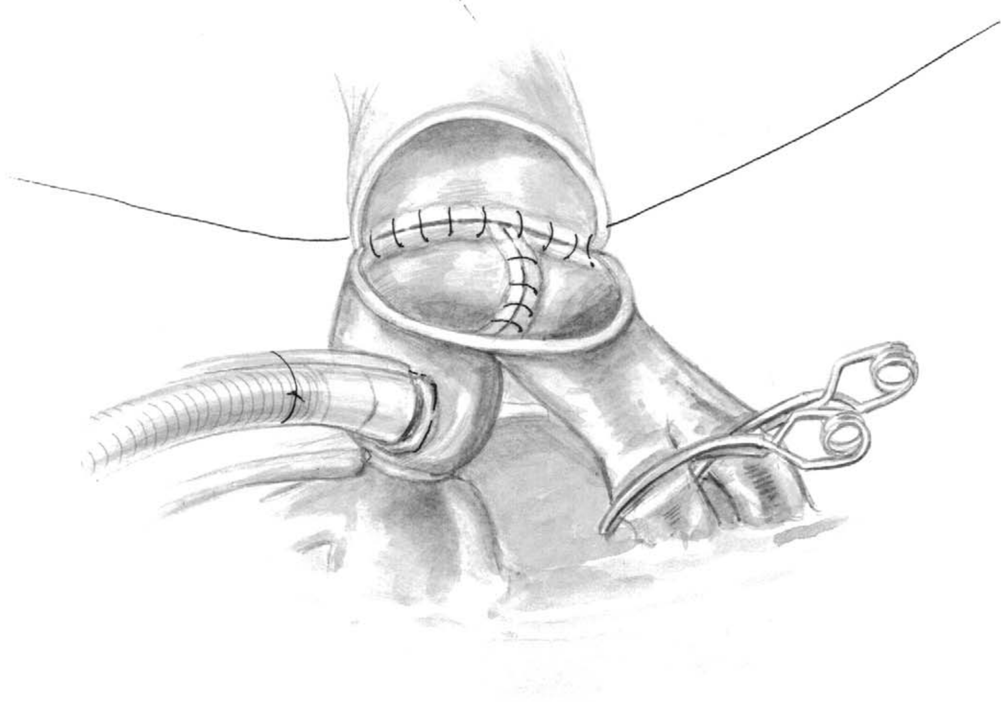



B

**Figure 14** This figure illustrates hepatic veins entering the atrium separate from the inferior vena cava (A). In this situation, the hepatic veins are removed from the atrium as a separate confluence from the cava (B). The veins are either controlled with neurosurgical (Codman) clips or allowed to drain into the pericardium with the return managed with a pump sucker. Adequate mobilization of the hepatic veins and inferior vena cava allows them to be joined side-to-side over a portion of their circumferences (C). The Gore-Tex ECC is then sewn to the amalgamated inferior cava and hepatic veins (D). vv = veins.




C



D

**Figure 14** (Continued)

## Postoperative Care

Postoperative care is similar to that following other cardiac operations in children. Particular considerations include the following. Postoperative monitoring includes central venous (Fontan) pressure measured through an internal jugular line, and common atrial pressure, measured via a transthoracic line. Every effort is made to extubate the patient soon after arrival in the ICU. To avoid fenestration and conduit thrombosis, intravenous heparin is initiated once bleeding has abated. The target partial thromboplastin time is 60 to 80 seconds. Once the central lines are removed, heparin is exchanged for aspirin, on which the patient is discharged. Warfarin has been used in patients with known hypercoagulability, or previous thromboses. Drainage tubes include a standard mediastinal chest tube, and Blake drains in the two pleural spaces. These are managed by a protocol in which they are removed once drainage is less than 2 mL/kg/d for each tube. It has been our experience that this protocol minimizes the need for chest tube reinsertion, or subsequent thoracentesis.

## Results

Several large series of ECC Fontan procedures have been reported. Marcelletti and coworkers reported 206 patients undergoing ECC from 1988 to 1998.[9] Operative mortality was 10%. There were no cases of conduit obstruction or thrombosis. Magnetic resonance imaging in 30 patients showed a reduction in conduit diameter of 18% during the first 6 months with no further change over the next 5 years. Tokunaga and colleagues reported 100 patients operated from 1994 to 2000 with no operative, and five late, deaths.[10] Petrossian and coworkers reported 256 patients operated from 1992 to 2004.[4] Operative mortality was 1%; 10-year freedom from mortality, takedown, or transplantation was 91%.

Several groups have reported concurrent comparisons of the ECC and LT approaches. From 1995 to 2006, 122 patients have undergone a Fontan procedure at MUSC. The approach was ECC in 75, and LT in 47 patients. The LT approach was primarily used earlier in the experience, and in patients with hypoplastic left heart syndrome.[11] Among the ECC procedures, 66 were primary Fontan procedures, and 9 were conversions of atriopulmonary Fontans. Operative mortality was 2.7% in the ECC group, and 2.1% in the LT group. In our experience, these two approaches have had comparable early and mid-term outcomes, including operative morbidity and mortality, postoperative hemodynamics, resource use, and mid-term survival and functional status.[11] Gaynor and coworkers and Azakie and coworkers similarly found that the type of Fontan procedure (ECC versus LT) was not a risk factor for either operative mortality or duration of pleural drainage.[7,12] Nakano and colleagues have reported similar exercise tolerance test results in 50 ECC and 67 LT patients at a mean of 5 to 6 years following surgery.[13]

One of the most important determinants of outcome after a Fontan procedure is the incidence of arrhythmias, both sinus node dysfunction and atrial tachyarrhythmias. There is compelling logic to the argument that the ECC should minimize, or avoid, arrhythmias. Nonetheless, early sinus node dysfunction (at hospital discharge) has been observed in 13 to 17% of patients undergoing an ECC Fontan procedure.[11,12,14,15] The incidence of sinus node dysfunction following a LT approach has been more variable. Some of this variation may be due to the approach taken to Fontan staging. A hemi-Fontan–LT approach can have an incidence of sinus node dysfunction as low as a bidirectional Glenn–ECC approach (0 to 13%).[11,14] In contrast, a bidirectional Glenn–LT approach has been reported to carry an incidence of early sinus node dysfunction of 23 to 46%.[12,15,16] This may be due to dissection, incisions, and suturing (at the time of LT) in the vicinity of the sinus node and artery, which may be obscured by scarring and absence of the superior cavo-atrial junction as a landmark.

The relative merits of the ECC and LT techniques may lie more in their respective rates of atrial tachyarrhythmias over the long term. Several studies have reported this incidence is lower after an ECC than a LT Fontan.[12,13,15] At present, the relative arrhythmogenic potential of the ECC and LT approaches is a question which remains incompletely answered. A more complete answer could be provided by prospective follow-up, including regularly scheduled Holter electrocardiograms, of patients undergoing consistent technical approaches to the two procedures.

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