Operative Techniques in Thoracic and Cardiovascular Surgery

Alternative Techniques for the Fontan Operation

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The surgical management of children with single ventricle physiology using a Fontan procedure has changed considerably over the past 3 decades. The introduction of the total cavopulmonary connection (TCPC) described by de Leval et al[1] was based partially on in vitro flow modeling. Subsequent studies with computational fluid dynamics (CFD)[2] have demonstrated designs of the TCPC connection that can cause flow disturbances and energy dissipation. Optimizing flow and diminishing power loss in the Fontan circuit presumably can improve hemodynamic efficiency and potentially improve long-term outcomes. Previous in vitro and CFD studies have shown reduced energy losses with caval offset[3] and with flaring of the cavopulmonary anastomosis,[4] thus prompting modification of the TCPC surgical procedure.

In 2007, using CFD, we proposed an optimized TCPC connection using a bifurcated Y-graft for the superior vena cava (SVC) to pulmonary artery connection and another bifurcated Y-graft for the inferior vena cava (IVC) to pulmonary artery connection[5] (U.S. Patent no. 7811244). Although this "Optiflo" connection had superior flow characteristics predicted by flow modeling, the optimized design from a surgical standpoint was cumbersome. Subsequent studies developing surgical designs to balance hepatic blood flow distribution to the right and left pulmonary arteries in children with acquired pulmonary arteriovenous malformations (AVMs) showed favorable results with a bifurcated Y-graft connection from the IVC to the branch pulmonary arteries.[6] Comparing the Y-graft Fontan connection with more conventional Fontan connections using CFD, others demonstrated improved flow characteristics, reduced energy losses, and balanced hepatic flow distribution (HFD) to the pulmonary arteries.[7,8] Based on these improvements in flow dynamics predicted by computerized modeling,[9,10] we have applied the Y-graft Fontan concept to children undergoing a Fontan procedure or a Fontan revision with a variety of anatomies.

Pulmonary Arteriovenous Malformations

Some children with an interrupted IVC with azygos continuation to the right SVC or hemiazygos continuation to a persistent left SVC undergo "definitive" palliation with a Kawashima operation.[11] This consists of a bidirectional cavopulmonary anastomosis (bilateral in the case of a persistent left SVC) without division of the azygos (hemiazygos) continuation. In effect, this routes all of the systemic venous return, except hepatic and mesenteric flow, to the pulmonary arteries. A significant percentage of these patients develop pulmonary AVMs in both lungs with resultant cyanosis.[12] This is analogous to the ipsilateral pulmonary AVMs observed with the classic Glenn anastomosis.[13] There seems to be some factor in the hepatic venous effluent which, if not circulated to the lung, results in the development of these AVMs. Rerouting the hepatic and mesenteric venous blood flow to the pulmonary arteries by completing the Fontan procedure can resolve this problem in most patients.[14-16]

Following the Kawashima procedure, completion of the TCPC was thus suggested as an efficient means to restore hepatic flow to the lungs and resolve pulmonary AVMs.[17] The TCPC does not completely eliminate the risk of pulmonary AVM development. Suboptimal design of the conduit from the hepatic veins may lead to an unbalanced HFD to the left and right lungs and in turn to unilateral pulmonary AVMs. We have shown that the low flow rate coming through the hepatic baffle in patients with an interrupted IVC (which accounts for only 20%-25% of the systemic venous return, as opposed to the normal 50%-60% carried by the IVC) increases the complexity of the flow interactions at the center of the TCPC, which in turn increases the difficulty of identifying the best surgical approach for a specific patient on the basis of anatomical considerations alone.[18] Thus, patients with an interrupted IVC remain at greater risk compared with other single ventricle cases for the development of pulmonary AVMs, even with a completed TCPC.

Our group recently presented a novel surgical planning framework[18] that enables one to virtually perform multiple surgical scenarios and determine the one that will yield the best hemodynamic performance before even entering the operating room. Such a surgical planning platform offers a unique solution for cases such as these, in which hemodynamics (and in particular, HFD) are vital to the operation but in which the small patient population and large number of anatomical variations pose a severe obstacle to the establishment of surgical guidelines from clinical studies alone.

Using computational flow dynamics, we have created a virtual surgical environment testing various patient-

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specific anatomies to determine the optimal surgical procedure for patients with interrupted IVC and severe pulmonary AVMs.[6] Figures 1 illustrates the application of the Y-graft Fontan procedure in a child with interrupted IVC with azygos continuation to the right SVC who had a Kawashima procedure. He developed pulmonary AVMs because of the lack of hepatic factor to the lungs so the Y-graft Fontan procedure was performed to distribute equal hepatic blood flow to both lungs (Figs. 2).

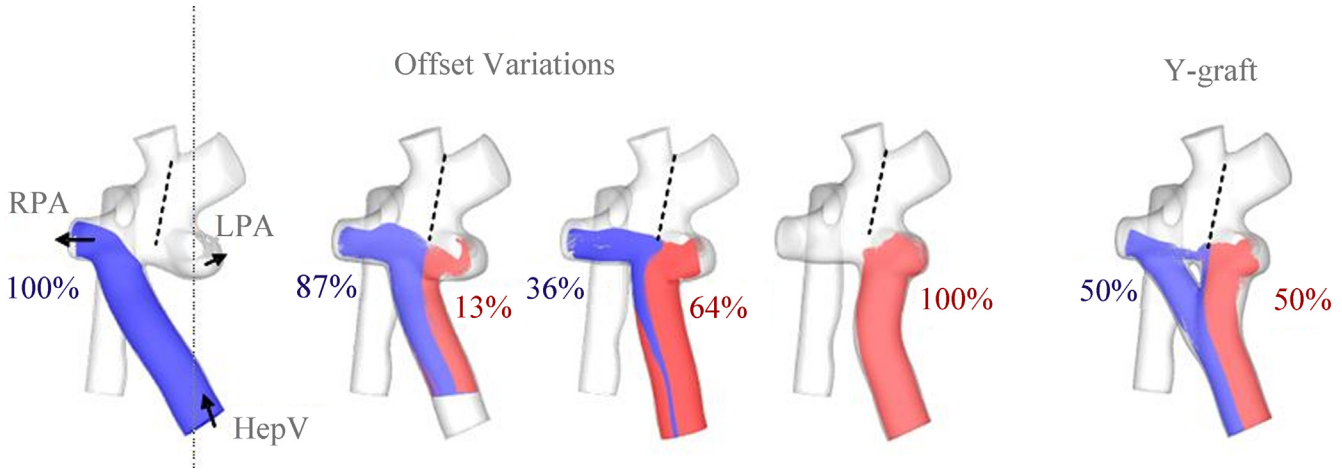

Figure 1 (A) Children with interrupted inferior vena cava with azygos continuation to the superior vena cava can be managed with a Kawashima procedure, which is effectively a bidirectional Glenn anastomosis. They can develop pulmonary arteriovenous malformations because of the lack of hepatic blood flow to the lungs. When this occurs, we have performed computational flow dynamic (CFD) studies of patient-specific MRI flows modeling the distribution of hepatic blood flow to the right and left pulmonary arteries exploring different surgical "solutions." The 4 images on the left demonstrate the extreme sensitivity of placement of a standard extracardiac Fontan graft resulting in differing distribution of hepatic flow to each lung (shown in percentages) varying greatly with minor differences in the site of anastomosis of the graft to the pulmonary artery. A more reliable solution is to direct hepatic blood flow to both lungs using a Y-graft Fontan procedure as shown on the right.

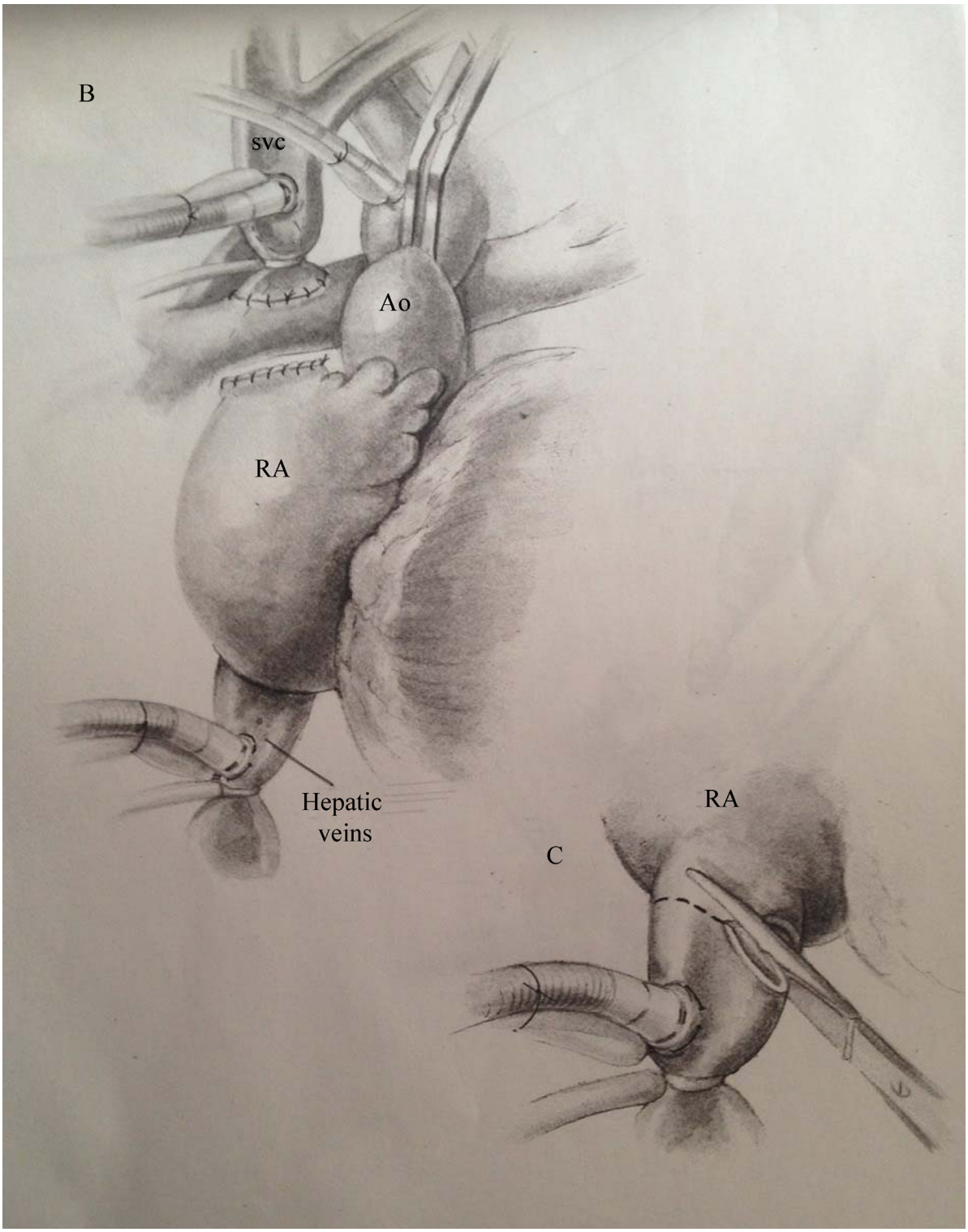

Figure 1 *(Continued)* (B) The cannulation strategy is aortobicaval cannulation, taking care to allow the interrupted inferior vena cava to drain to the superior vena caval cannula by appropriately positioning the superior vena caval tourniquet. (C) After cardioplegic arrest, the hepatic venous confluence is transected off the bottom of the right atrium. Ao = aorta; RA = right atrium.

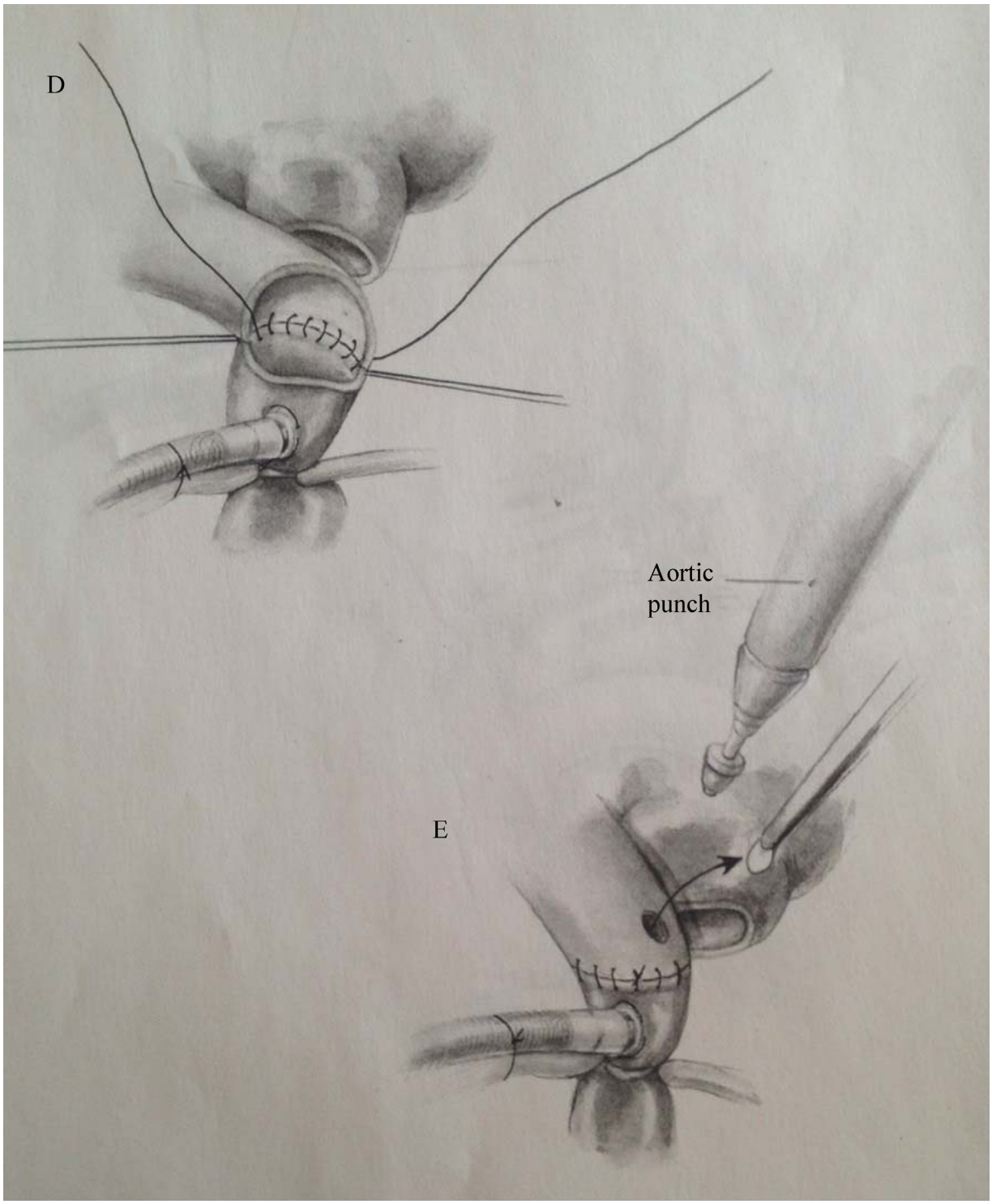

Figure 1 *(Continued)* (D) A 18 or 20-mm portion of a commercially available bifurcated aortoiliac 18 × 9 × 9 or 20 × 10 × 10 expanded polytetrafluoroethylene (PTFE) graft is then anastomosed end-to-end to the hepatic venous confluence with continuous polypropylene suture. (E) We routinely create a fenestration using either a 2.8 or 4.0-mm aortic punch in the PTFE graft 5-10 mm cephalad to the suture line between the hepatic venous confluence and the PTFE graft.

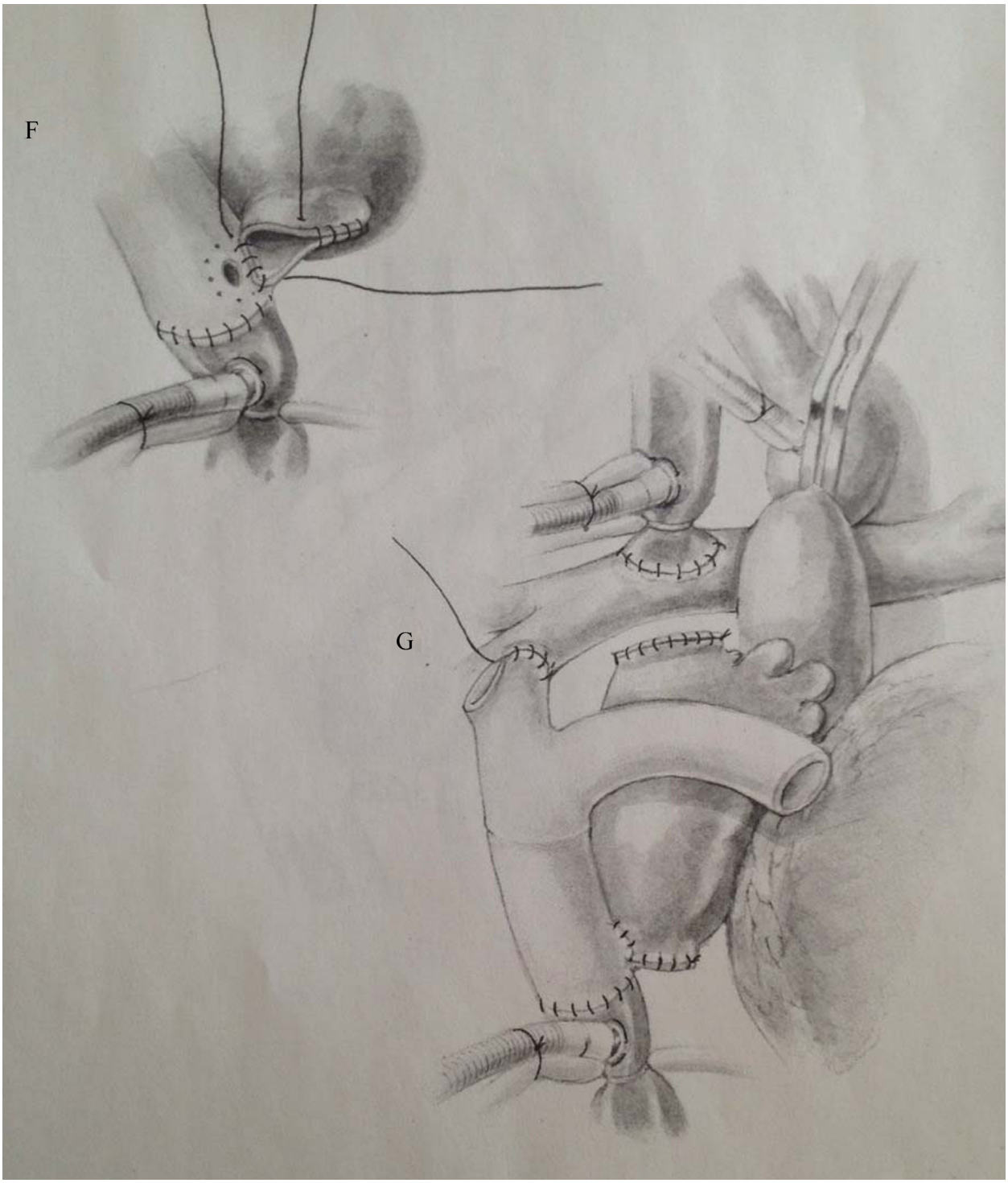

Figure 1 *(Continued)* (F) The medial aspect of the floor of the right atrium from which the hepatic venous confluence was divided is partially closed for half of its length. The lateral portion of the right atrial incision is sewn around the fenestration in the PTFE graft with shallow bites into the graft away from the fenestration to allow for unimpeded flow through the fenestration postoperatively. (G) The right limb of the bifurcated portion of the PTFE Y-graft is anastomosed to the undersurface of the right pulmonary artery. It is important to direct the right limb of the graft away from the bidirectional Glenn connection to avoid turbulent flow resulting in unfavorable energetics. To achieve this, we attempt to anastomose the right limb of the graft onto the right pulmonary artery after the take off of the right upper lobe artery.

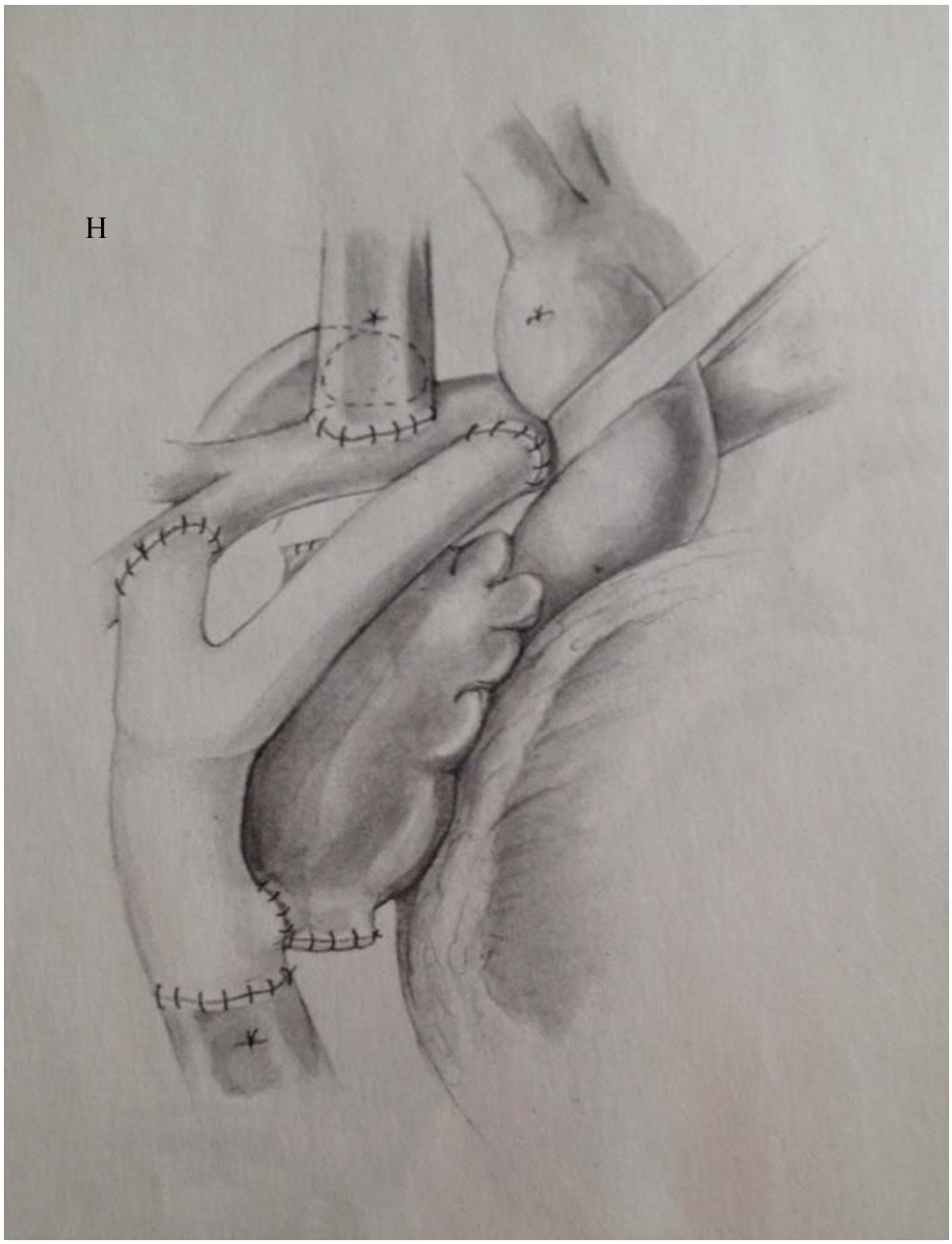

Figure 1 *(Continued)* (H) The left limb of the Y-graft is anastomosed to the proximal left pulmonary artery behind the aorta in an oblique fashion, also away from the Glenn anastomosis to avoid turbulence. This picture depicts the final operation with the fenestration to the bottom of the right atrium and the Y-graft to the right and left pulmonary arteries.

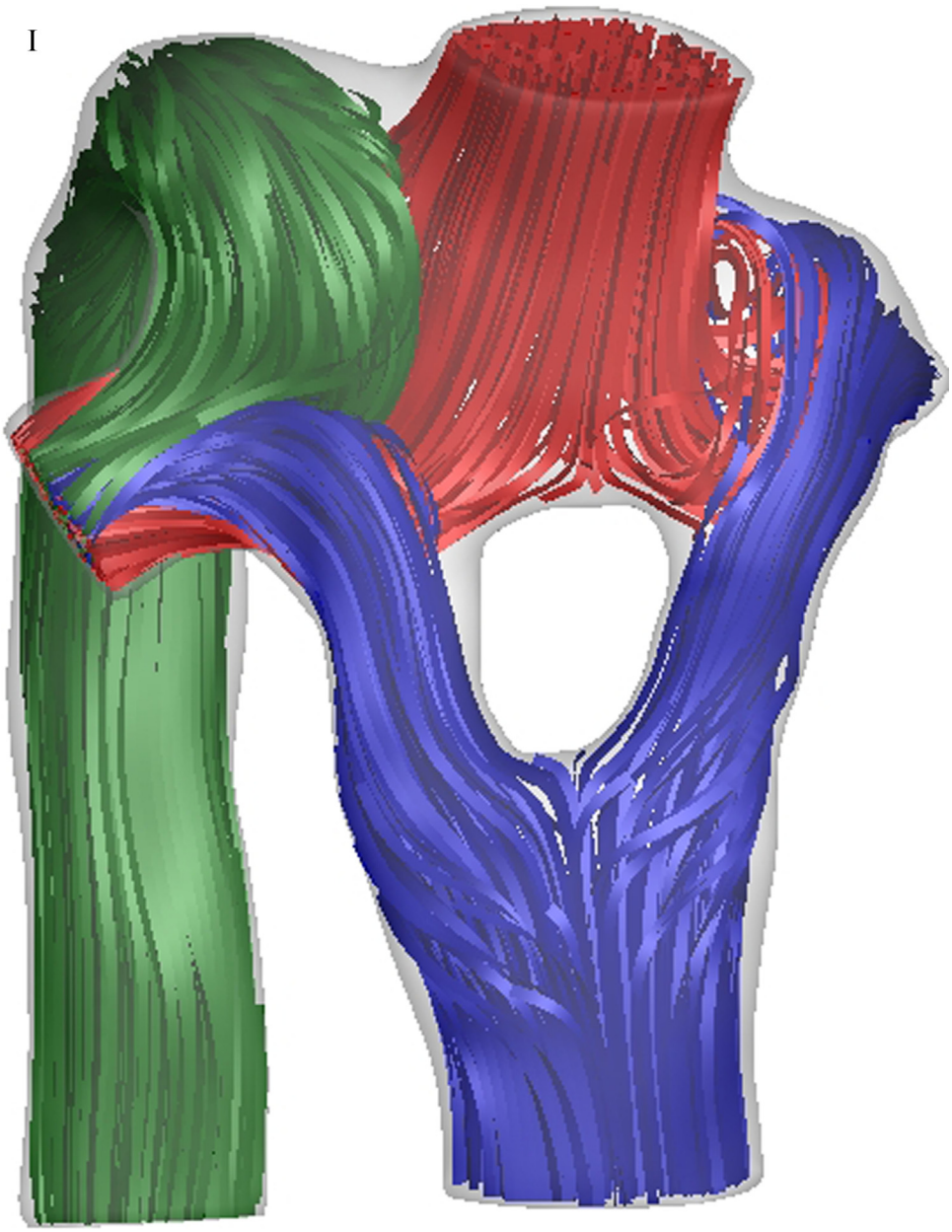

Figure 1 *(Continued)* (I) A computational flow model (CFD) of a completed Y-graft Fontan shows near laminar flow from the Y-graft to the branch right and left pulmonary arteries with balanced hepatic flow distribution to both the right and left pulmonary arteries. (Color version of figure is available online.)

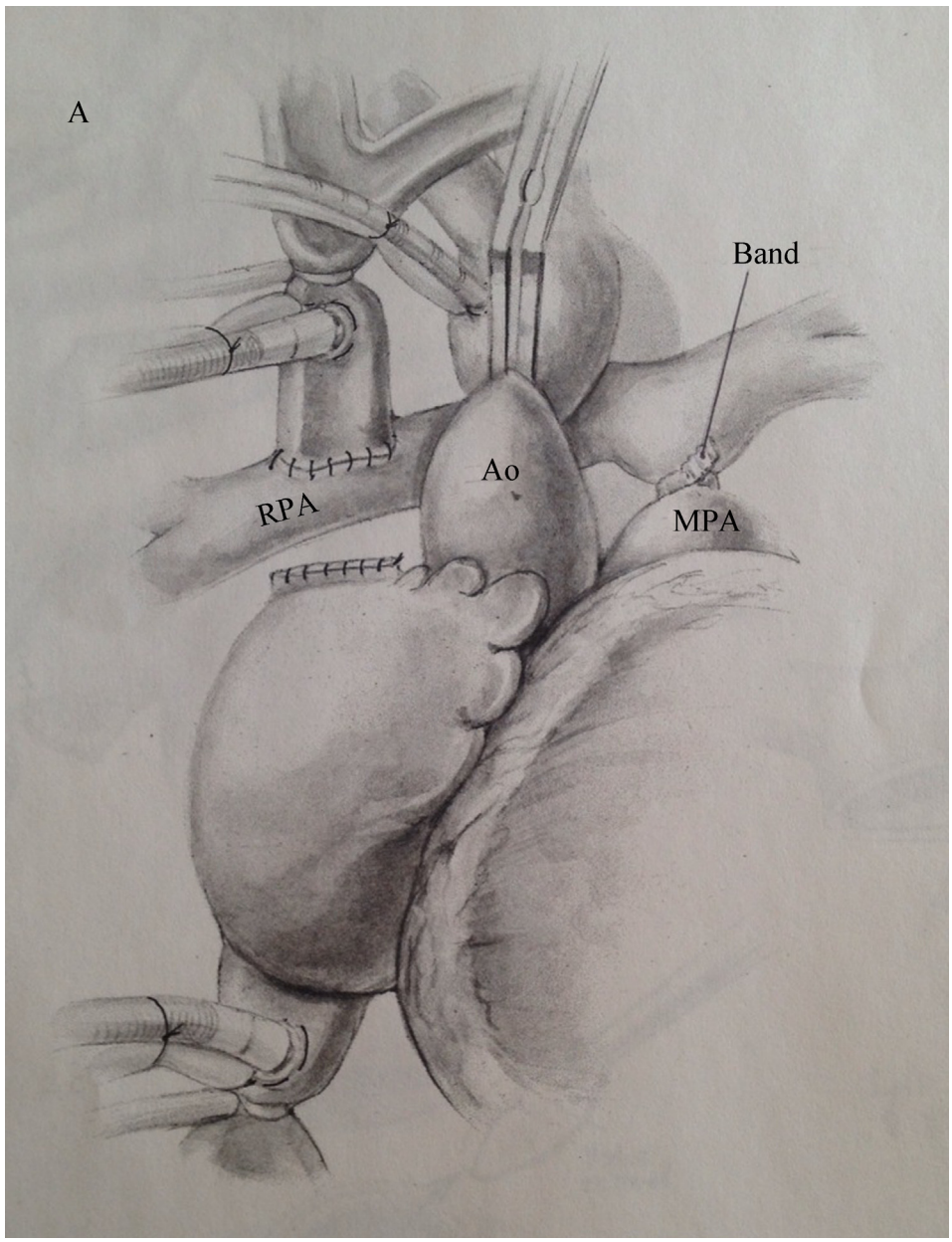

Figure 2 (A) The Y-graft concept can also be applied to more conventional anatomy as shown in this patient who previously underwent a bidirectional Glenn anastomosis with a very tight pulmonary artery band. The cannulation technique is aortobicaval cannulation. RPA = right pulmonary artery; Ao = aorta; MPA = main pulmonary artery.

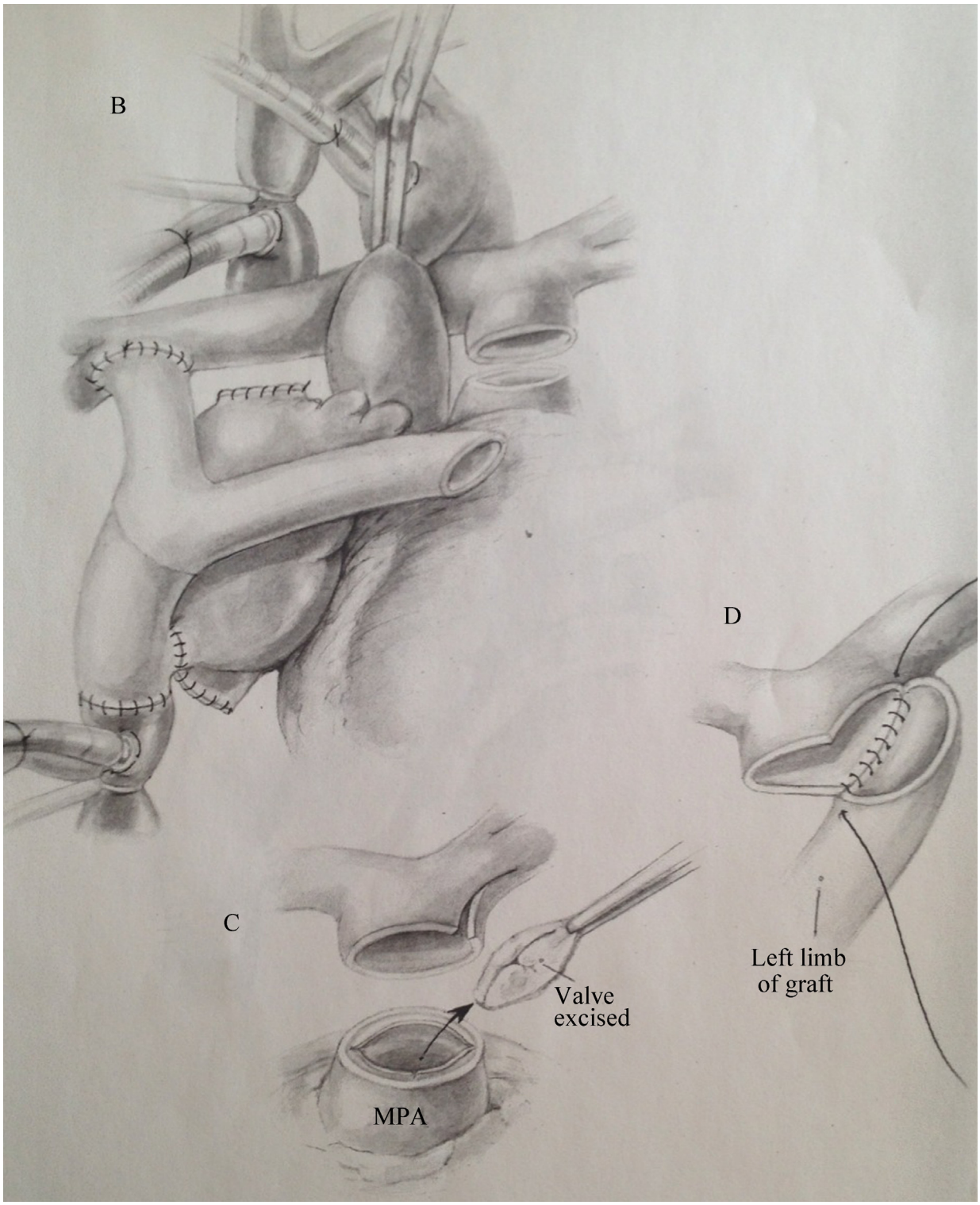

Figure 2 *(Continued)* (B) After establishing cardiopulmonary bypass and cardioplegic arrest, the inferior portion of the PTFE Y-graft is performed as shown in Figures 1a,1bc,1de,1fg,1h,1i, as is the right limb of the graft. For patients who have had previous pulmonary artery banding, we have found it useful to remove the pulmonary artery band and extend the incision of the distal main pulmonary artery segment onto the proximal left pulmonary artery for the left limb of the Y-graft. (C) The pulmonary valve is excised and the proximal pulmonary trunk is oversewn to prevent a blind pouch with the potential for development of clots. (D) A generous oblique anastomosis between the left limb of the Y-graft and the distal main pulmonary segment extending out onto the left pulmonary artery is shown. This routes the left limb of the Y-graft behind the ascending aorta. MPA = main pulmonary artery.

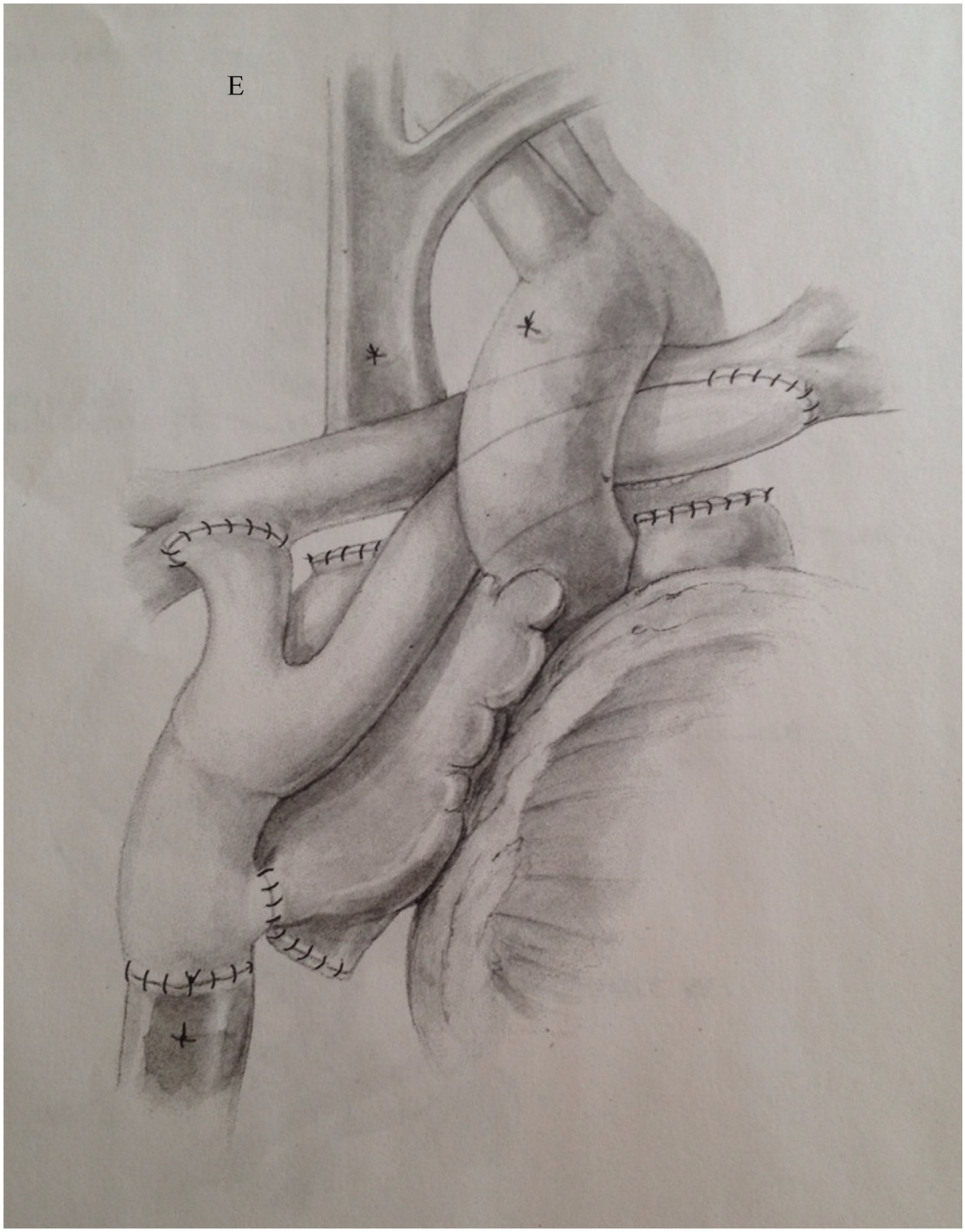

Figure 2 *(Continued)* (E) The completed Y-graft Fontan is shown, again emphasizing the importance of anastomosing the right and left limbs of the Y-graft away from the Glenn anastomosis. The fenestration to the right atrium is also depicted.

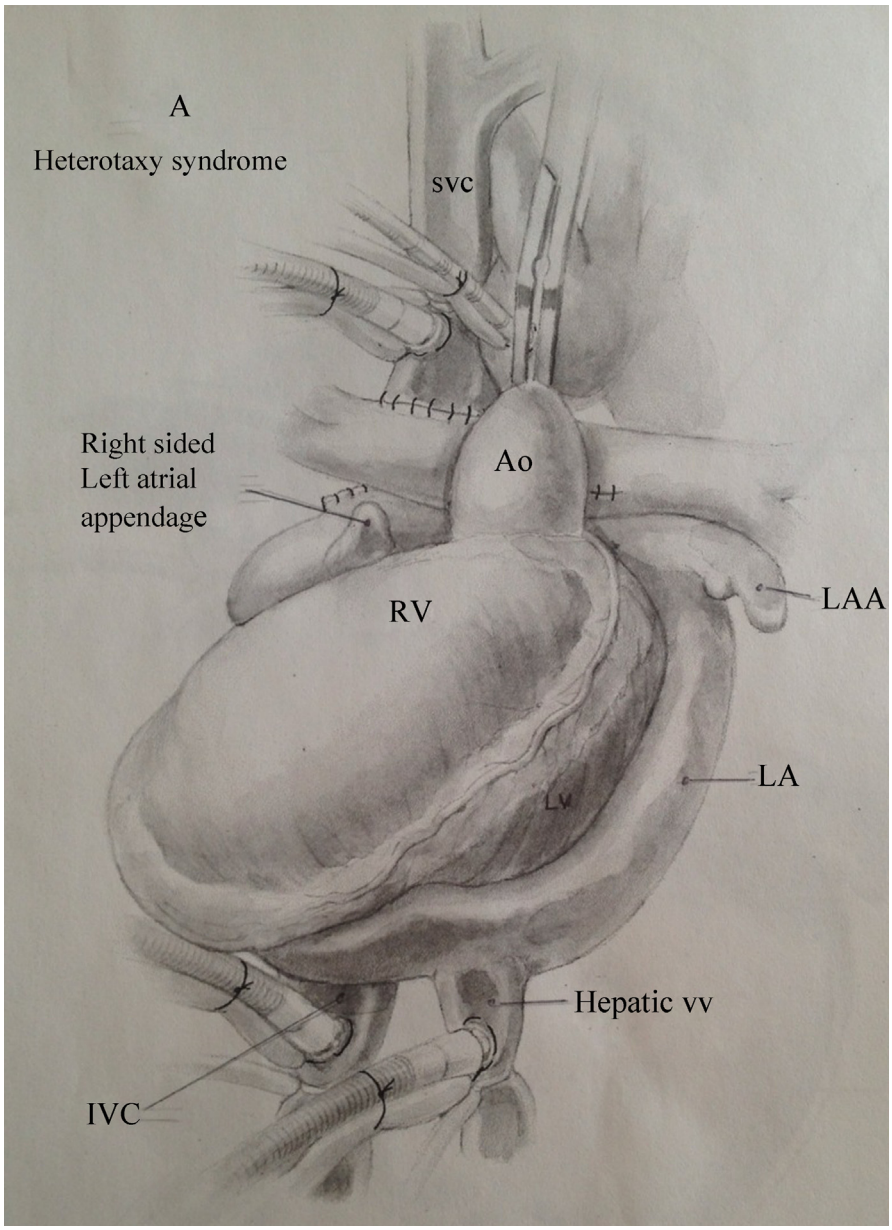

Figure 3 (A) Patients with heterotaxy syndrome can present with very challenging anatomies that make an extracardiac Fontan particularly difficult. An example is shown here in a patient with situs solitus, left atrial isomerism, and effectively, a common atrium, who had a previous bidirectional Glenn anastomosis. There is separate insertion of the hepatic veins into the floor of the common atrium in the midline in addition to the inferior vena cava coming up in the usual location. The dextrocardia makes an extracardiac Fontan more difficult; therefore, we choose to create an intra-atrial baffle in these patients. The patient is cannulated with a cannula in the ascending aorta for arterial inflow and venous return through a superior vena caval cannula and separate cannulas in the inferior vena cava and the hepatic venous confluence. SVC = superior vena cava; Ao = aorta; RV = right ventricle; LAA = left atrial appendage; LV = left ventricle; LA = left atrium; IVC = inferior vena cava.

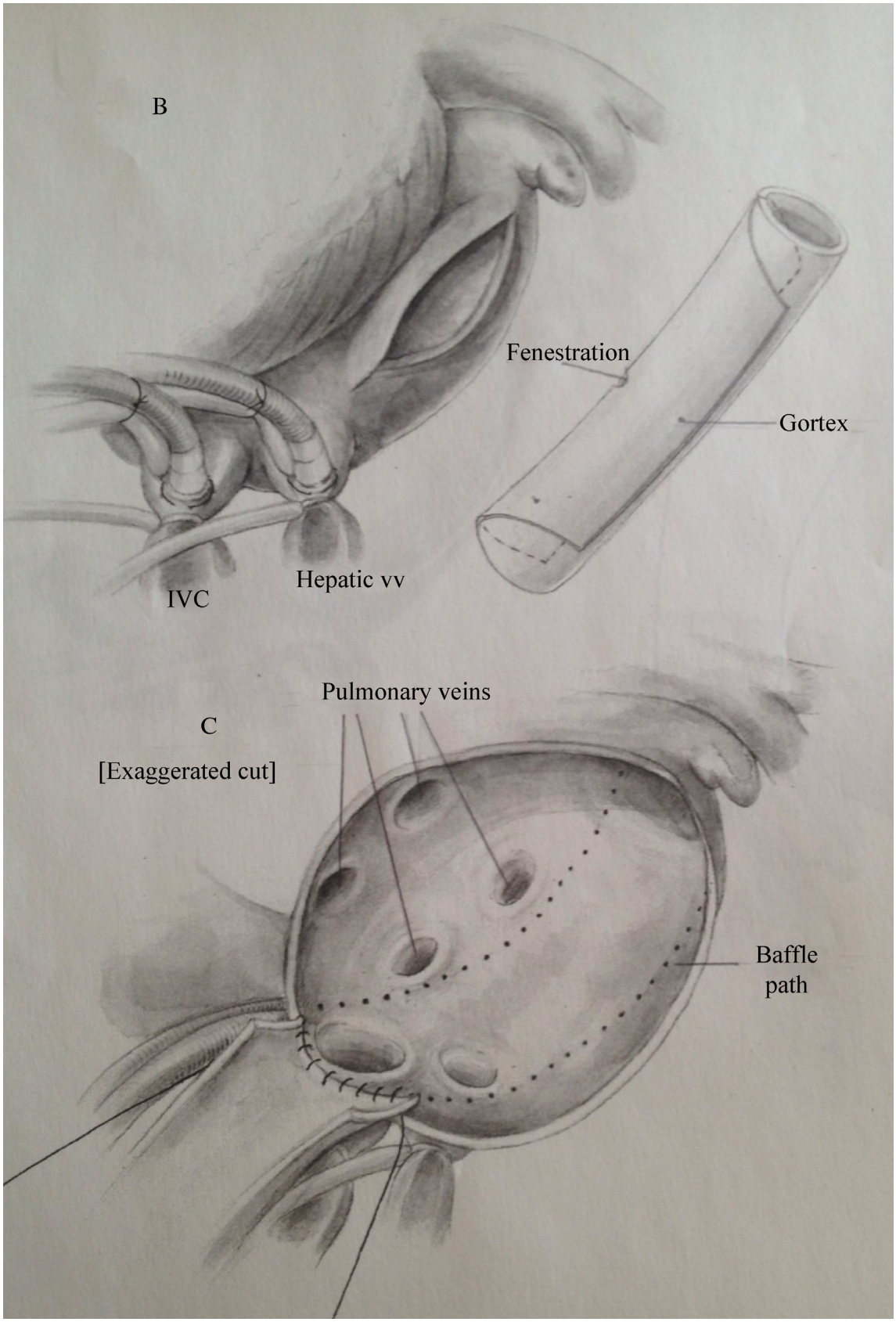

Figure 3 *(Continued)* (B) After cardioplegic arrest, the heart is rotated over to the right, giving access to the left lateral aspect of the common atrium. An incision is made on the left lateral aspect of the common atrium. A 14 or 16-mm PTFE graft is opened longitudinally, so about half of its circumference is available. We typically would create a single fenestration with a 2.8 or 4.0-mm aortic punch. (C) With opening the left lateral aspect of the common atrium, one can easily visualize the orifices of the inferior vena cava and the hepatic veins entering separately. The pulmonary veins tpically come in on either side of the midline. The dotted line depicts the suture line of the intra-atrial baffle channeling both the inferior vena cava and hepatic venous return up to the base of the left-sided left atrial appendage. IVC = inferior vena cava.

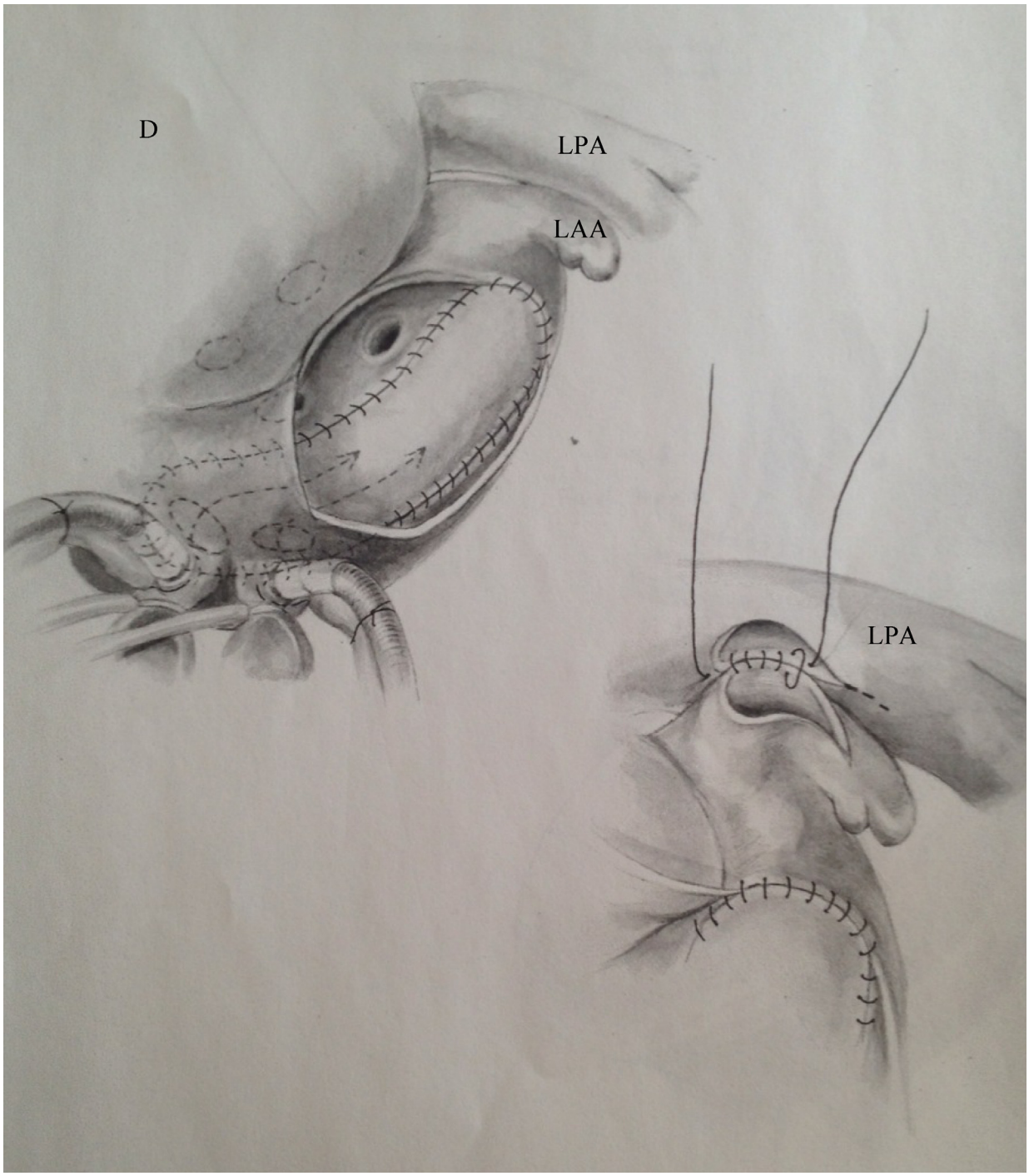

Figure 3 *(Continued)* (D) The completed intra-atrial baffle is shown, with the superior aspect of the intra-atrial baffle sewn to the base of the left atrium with continuous suture. This results in a cylindrical baffle channeling the inferior vena cava and hepatic venous confluence to the base of the left atrium, in which about half of the cylinder is PTFE material and the remaining half is native atrial tissue. The inset shows the undersurface of the proximal left pulmonary artery opened and the superior aspect of the left-sided left atrial appendage anastomosed to the undersurface of the left pulmonary artery channeling the intra-atrial baffle to the left pulmonary artery. LPA = left pulmonary artery; LAA = left atrial appendage.

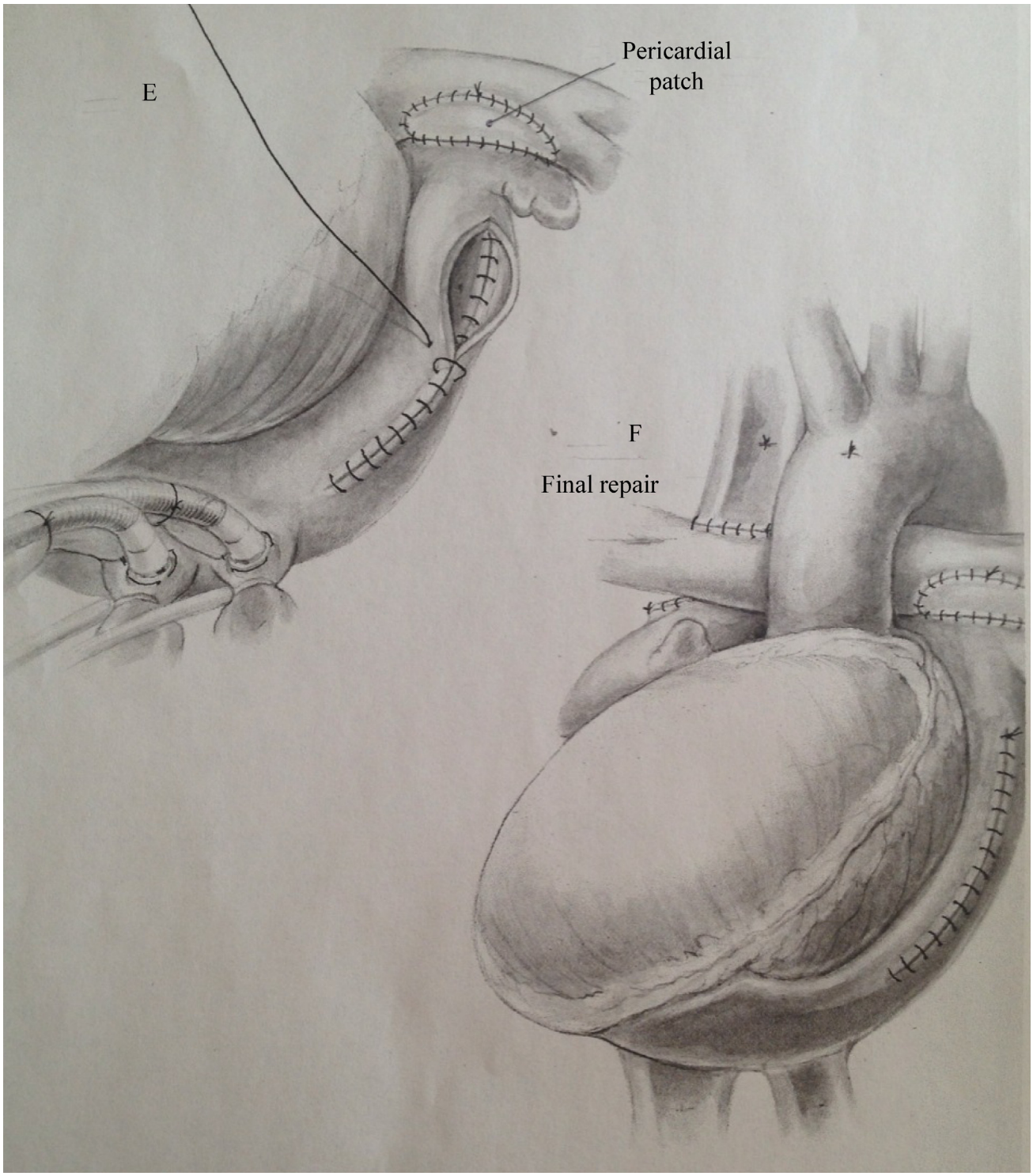

Figure 3 *(Continued)* (E) The anterior wall of the anastomosis between the left atrial appendage and the left pulmonary artery is augmented with a patch of autologous pericardium or homograft material. The atriotomy is closed primarily. (F) The completed repair shows a fenestrated lateral tunnel of Fontan, successfully completing the Fontan procedure in this patient with heterotaxy syndrome and left atrial isomerism. LPA = left pulmonary artery.

Postoperative Care and Results

The postoperative management of patients undergoing a Y-graft Fontan procedure or a complex intra-atrial tunnel (Figs. 3a,3bc,3d,3ef) for the patient with heterotaxy syndrome is no different from that for a patient having a more standard extracardiac or lateral tunnel Fontan procedure. We strive for early extubation within the first few hours after arrival in the intensive care unit. Since August 2010, 34 patients have had a Y-graft Fontan at our institution without mortality. The average intensive care unit stay is 1.8 days and the average hospital stay is 7.9 days—all patients stay hospitalized until all chest tubes are removed. All patients receive 81 mg of aspirin daily postoperatively. We do not use other forms of anticoagulation. To date, no patient with a Y-graft Fontan has required reoperation, although 2 patients have had intervention in the cardiac catheterization laboratory for distal left pulmonary artery stenosis. No patient has had device closure of the fenestration.

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