Operative Techniques in Thoracic and Cardiovascular Surgery

The Norwood Procedure with an Innominate Artery-to-Pulmonary Artery Shunt

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Norwood and colleagues reported the first successful palliation of hypoplastic left heart syndrome (HLHS) in 1983.[1] Today, there are several competing surgical strategies for stage I palliation, including the right-ventricle-to-pulmonary-artery conduit (Sano) modification, the so-called hybrid procedure (ductal stenting and branch pulmonary artery banding), and perhaps the gold standard, the Norwood procedure with an innominate artery-to-pulmonary artery (Blalock-Taussig) shunt.[2-4] Regardless of the surgical approach, neonatal palliation of hypoplastic left heart requires relief of systemic outflow obstruction, provision of unobstructed coronary blood flow, creation of a nonrestrictive atrial septal communication, and limitation of excessive pulmonary blood flow. Furthermore, a successful program for management of the patient with HLHS must include strategies to ensure a stable postoperative course and minimal interstage loss and should aim for optimal late neurodevelopmental outcome. This article will detail the Norwood procedure with an innominate artery-to-pulmonary artery shunt as performed at the Children's Hospital of Wisconsin. Like most centers, our approach has changed over time and continues to evolve. Whether or not the Sano modification supplants the modified Blalock-Taussig shunt remains to be determined; nevertheless, many of the issues are common to both procedures, and even advocates for the Sano modification may find some of this description useful.

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Description of Procedure

Preoperative stabilization is essential. Increasingly, patients are diagnosed antenatally, and supportive measures are undertaken immediately after delivery. For patients presenting in the newborn period either with impending ductal closure or heart failure, a period of resuscitation will be necessary. Except in the rare patient who is unresponsive to prostaglandin E1 or who has a critically restrictive atrial septal defect, surgery should not take place until circulation has been stabilized and low output/acidosis resolved. Surgery is typically scheduled electively about 2 to 5 days after presentation. We routinely use preoperative steroids given in 2 doses (30 mg/kg of solumedrol intravenously each), 9 hours and 3 hours before skin incision. Aprotinin (Bayer Corp, Leverkusen, Germany) is used in all patients. Initial arterial cannulation is via a Gore-Tex graft (WL Gore Co., Newark, DE) anastomosed end-to-side to the innominate artery. This graft will ultimately become the innominate artery-to-pulmonary artery shunt. Phenoxybenzamine (.25 mg/kg) is added to the cardiopulmonary bypass circuit with initiation of cardiopulmonary bypass. We cool to a bladder temperature of 18°C to 20°C over at least 30 minutes with a pH-stat perfusion strategy for cooling. The hematocrit is maintained above 25%. We routinely use continuous cerebral perfusion to minimize the period of deep hypothermic circulatory arrest. Flow is maintained to the innominate artery at a rate of 20 to 30 mL/kg/min, and near-infrared spectroscopy is used to assist in achieving adequate perfusion. Modified ultrafiltration is used routinely after separation from cardiopulmonary bypass. Postoperative management is facilitated by the routine use of an oximetric catheter (Abbot Laboratories, North Chicago, IL) and 2-site near-infrared spectroscopy (Somanetics Corp, Troy, MI) that has been previously described.[5,6] The steps in stage I reconstruction are detailed in Figures 1 to 13.

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Surgical Procedure

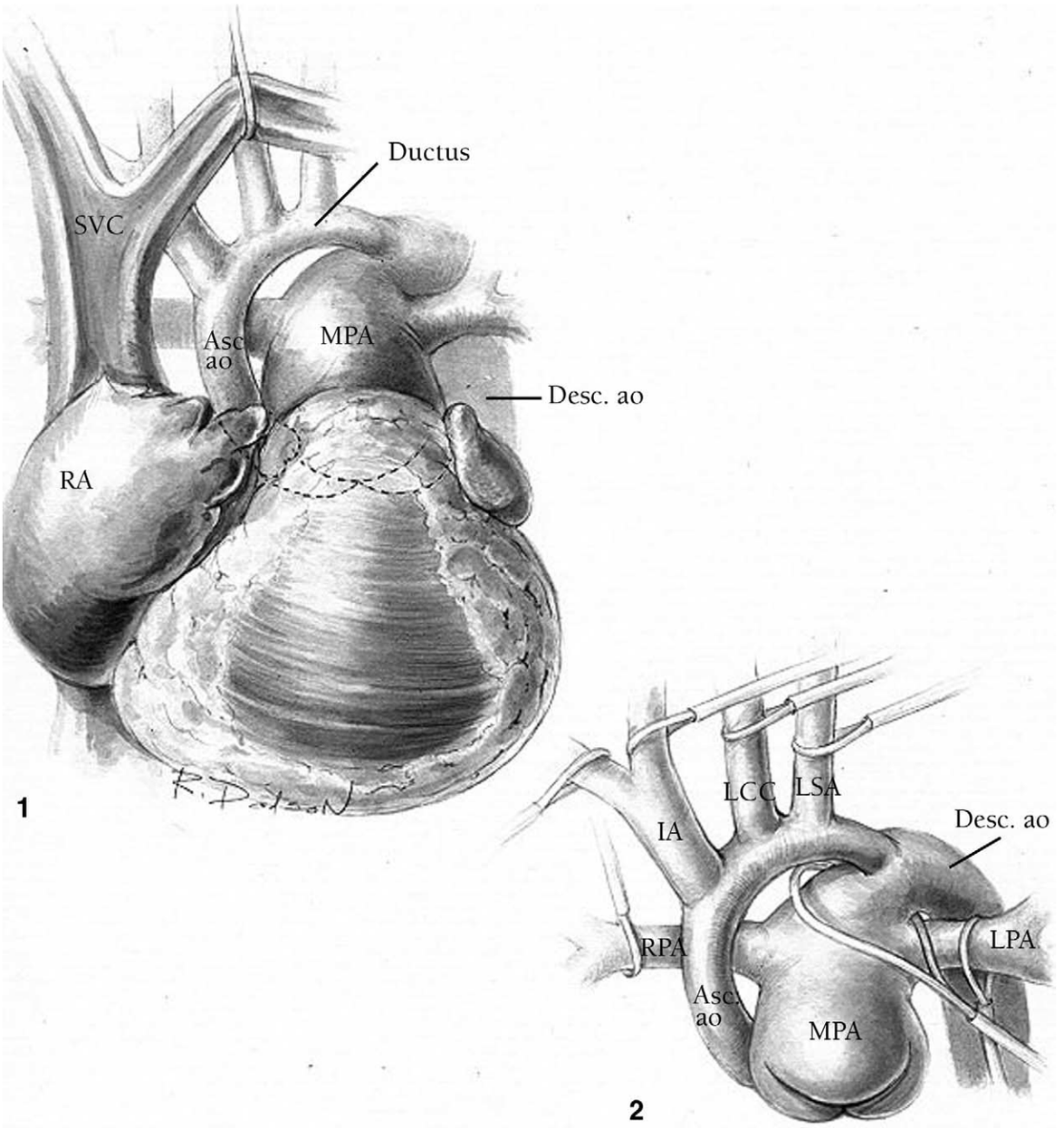

Figure 1 After a median sternotomy, the pericardium is opened and the vascular structures are mobilized. The innominate vein is mobilized and looped with a tape; this allows for atraumatic retraction and facilitates exposure of the brachiocephalic vessels.

Figure 2 The brachiocephalic vessels are fully mobilized. The ascending aorta is separated from the main pulmonary artery, and the arch and ductus arteriosus are also mobilized. The branch pulmonary arteries are mobilized to their first branches, similar to the degree of mobilization used for an arterial switch.

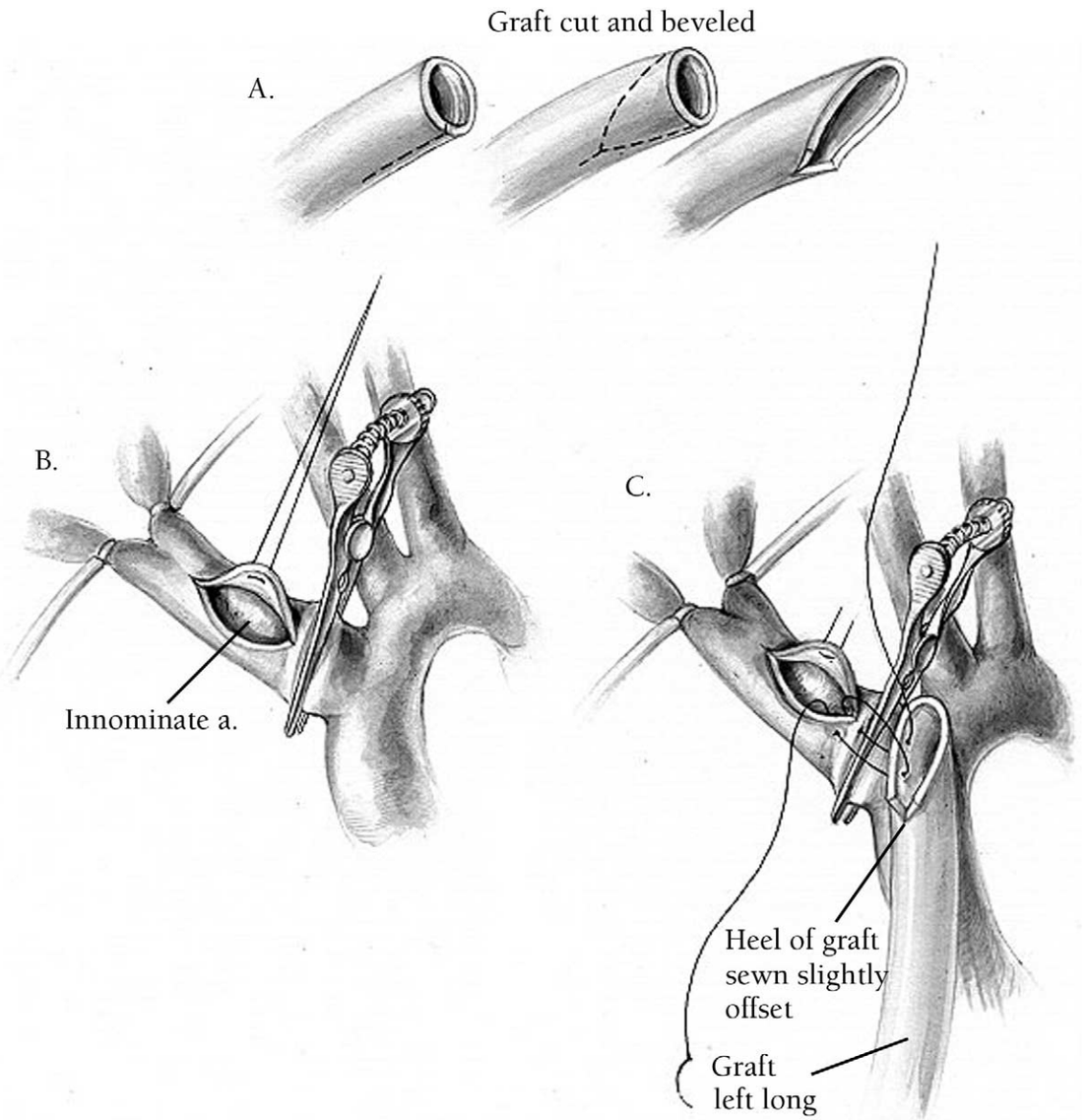

Figure 3 The innominate artery is isolated by placing a bulldog clamp across its origin and tightening snares around the right subclavian artery and right carotid artery. A Gore-Tex graft, destined to become the systemic-to-pulmonary artery shunt but used initially for arterial cannulation, is brought up and cut in a beveled manner as shown. This results in a large "cobra head" connection between the proximal graft and the innominate artery. The caliber of the graft is dependent on the size of the patient. For patients under 3.5 kg, we generally use a 3.0-mm graft, and for patients greater than 3.5 kg, we commonly use a 3.5-mm graft. Rarely, larger grafts are used for larger patients or those who present late (>2 weeks of age). The anastomosis of the graft to the innominate artery is slightly offset to achieve a more vertical orientation of the innominate artery-to-pulmonary artery shunt.

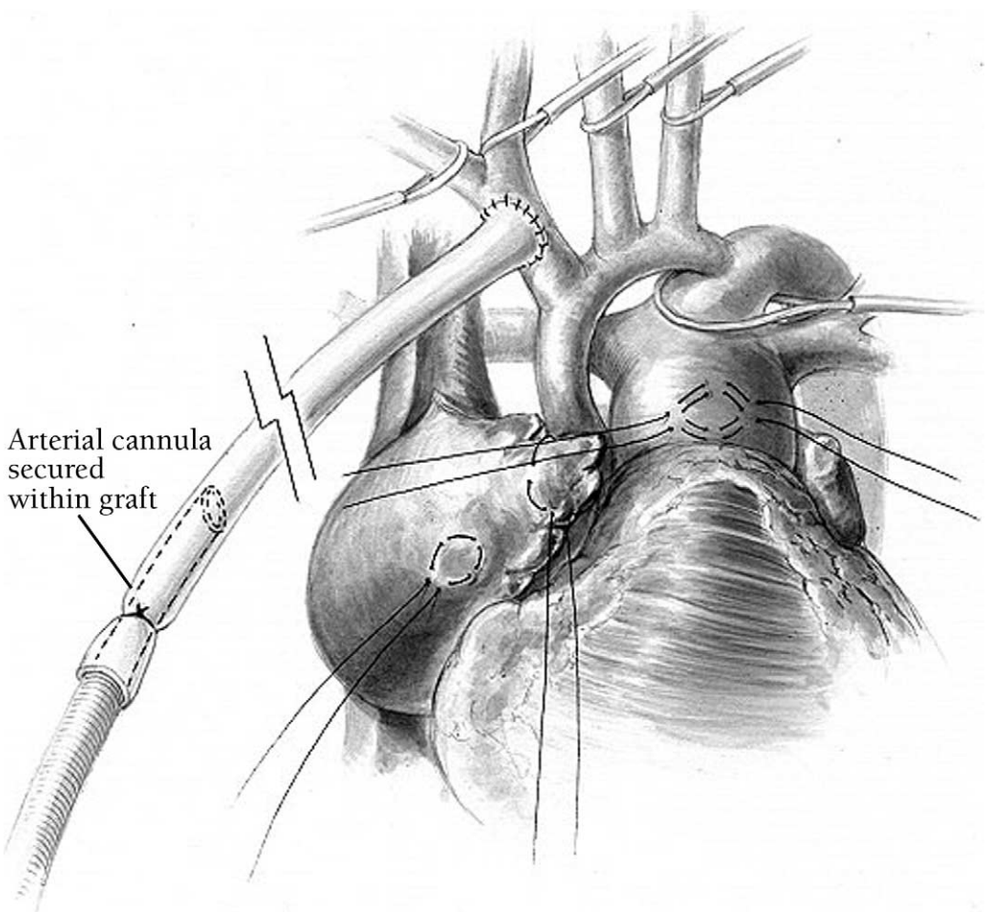

Figure 4 The Gore-Tex graft is used for arterial cannulation; an 8F arterial cannula (Medtronic, Minneapolis, MN) is used. Additional pursestrings of 5-0 prolene (Johnson and Johnson, New Brunswick, NJ) are placed. A double pursestring is placed in the proximal pulmonary artery just above the anterior commissure of the pulmonary valve; this will be used later for neoaortic cannulation. A pursestring is placed in the right atrial appendage for venous cannulation, and a pursestring is placed in the right atrial freewall for a vent that will be placed in the right ventricle.

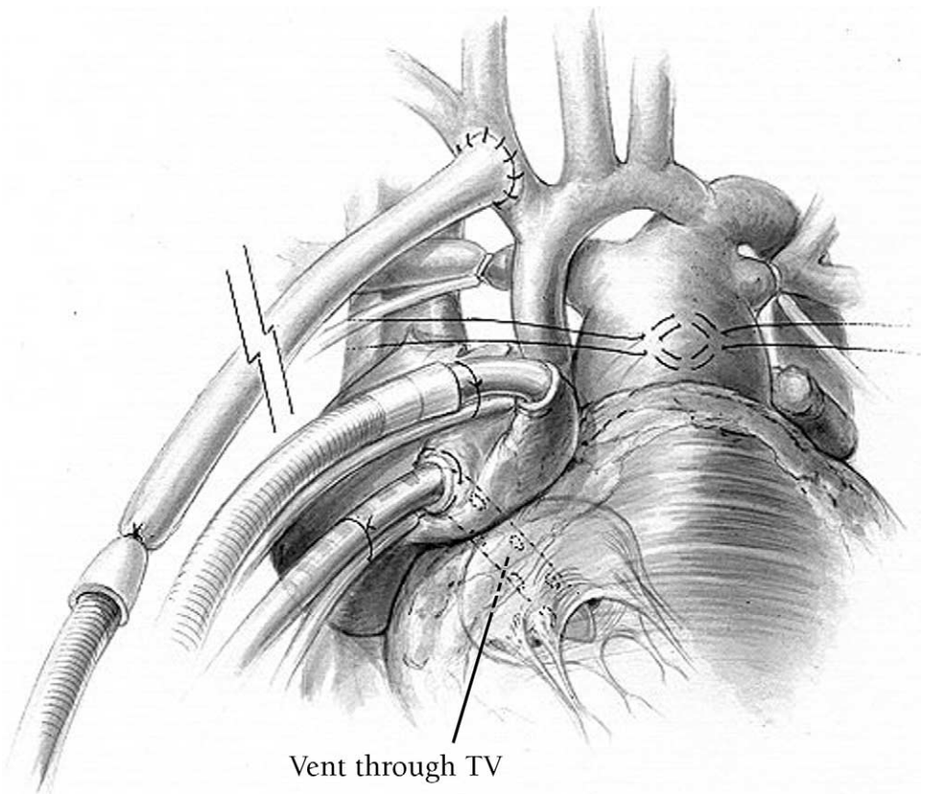

Figure 5 The right atrial appendage is cannulated with a 16F venous cannula (Medtronic). Cardiopulmonary bypass is instituted, and the patient is cooled over 30 minutes to a bladder temperature of less than 20°C. With initiation of cardiopulmonary bypass, the branch pulmonary arteries are snared, and a 13F ventricular vent (Medtronic) is placed through a separate pursestring in the right atrial freewall and directed across the tricuspid valve into the right ventricle. The vent is useful in preventing ventricular distension as contractions become less effective with hypothermia.

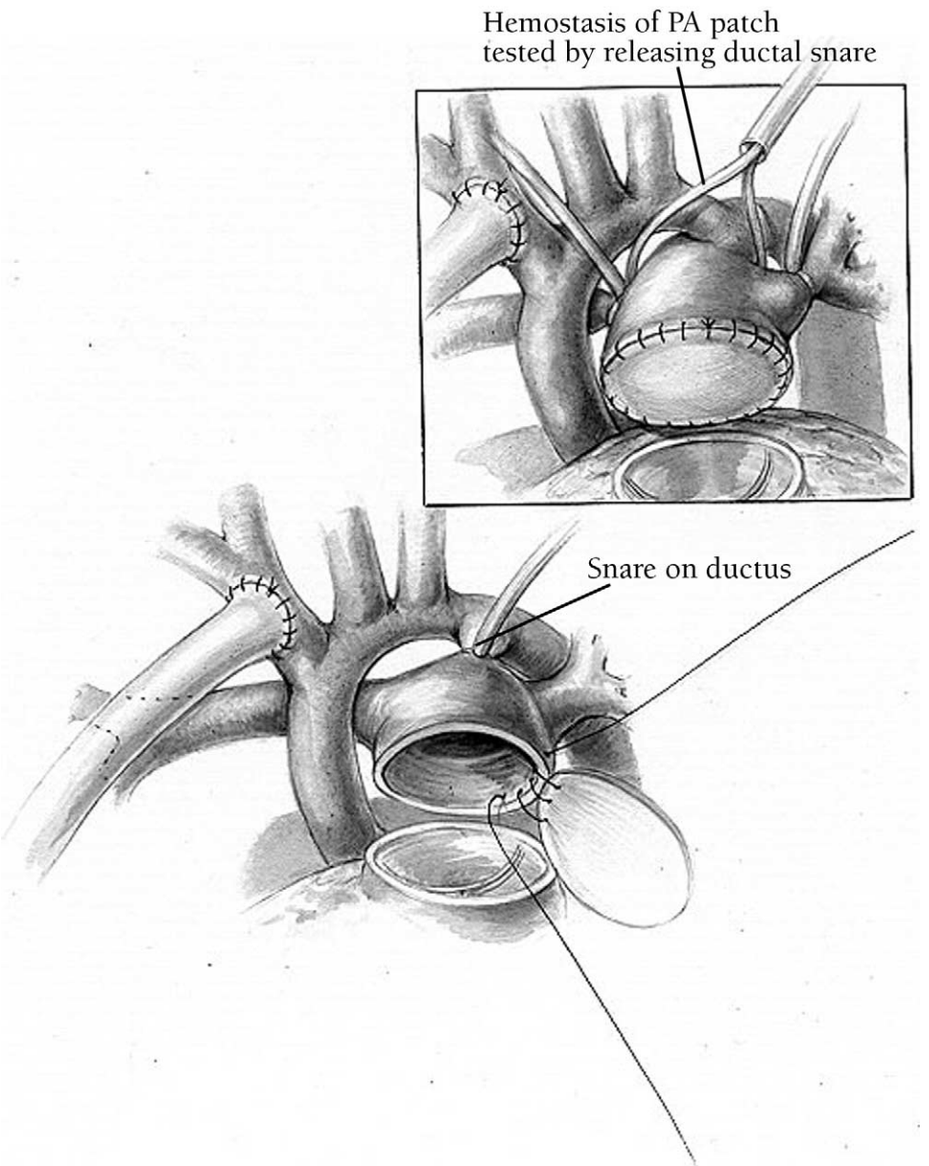

Figure 6 After cooling for approximately 10 minutes, the ductus is snared, and the main pulmonary is divided just proximal to the takeoff of the branch pulmonary arteries. Although opening of the pulmonary artery can result in entrainment of air into the venous cannula, the vent is more than capable of handling the venous return. Alternatively, the proximal pulmonary artery can be controlled with a Cooley carotid clamp (Fehling Surgical Instruments, Acworth, GA). The distal pulmonary artery confluence is patched with pulmonary homograft. Reconstruction is performed with 7-0 Prolene suture. After completing this patch, hemostasis of the suture line can be tested by snaring the branch pulmonary arteries and releasing the snare on the ductus as shown in the inset. Any bleeding points are reinforced with additional sutures. Careful attention to suture placement and hemostasis throughout the procedure will decrease the potential for bleeding after separation from bypass and improve postoperative stability.

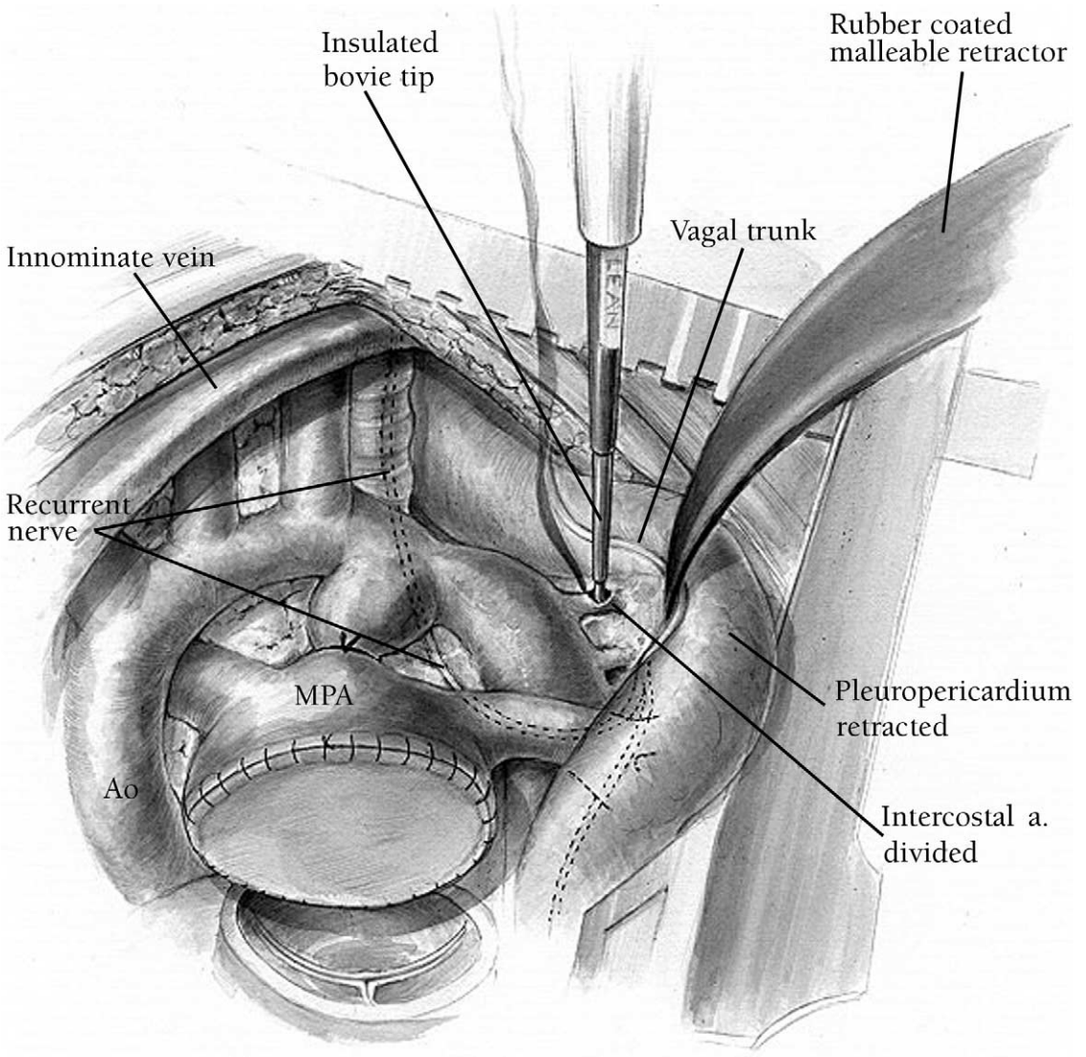

Figure 7 Mobilization of the proximal descending thoracic aorta. After the pulmonary artery confluence is patched, the ductus arteriosus is ligated, and the proximal descending aorta is mobilized during the final 10 minutes of cooling. An insulated malleable retractor is used to retract the pericardium and pleura. An insulated cautery is used at low settings (10%) to divide the first 2 or 3 sets of intercostal branches. The use of an insulated retractor and cautery tip minimizes the risk of recurrent nerve injury, which can be the source of respiratory and feeding difficulties.

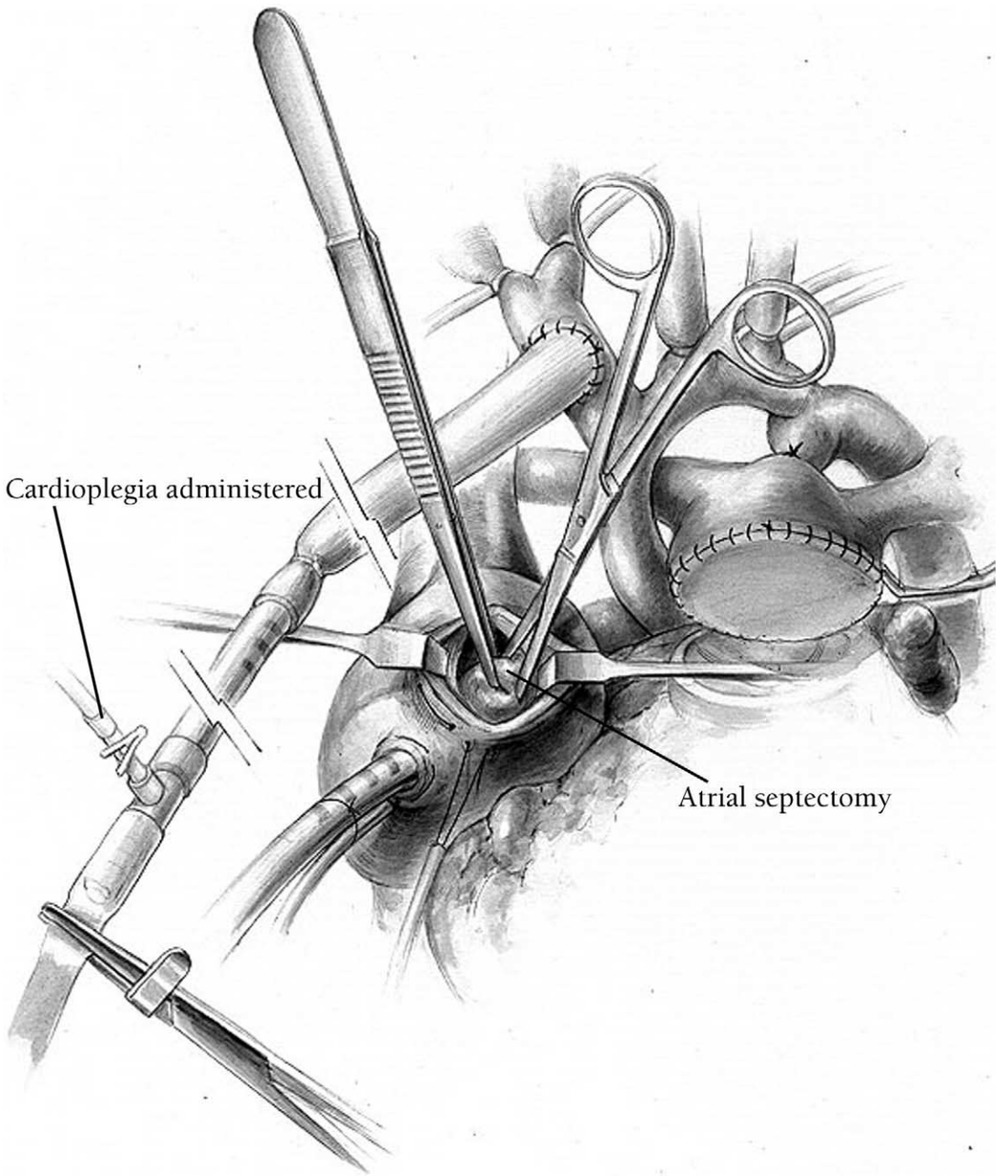

Figure 8 Delivery of cardioplegia and atrial septectomy. After cooling for at least 30 minutes and reaching a bladder temperature between 18°C and 20°C, circulatory arrest is established. The arterial line is clamped, the brachiocephalic vessels are snared, and the descending thoracic aorta is clamped. Cardioplegia is delivered via a side arm on the arterial cannula. While cardioplegia is being delivered, the venous cannula is removed and an atrial septectomy is performed.

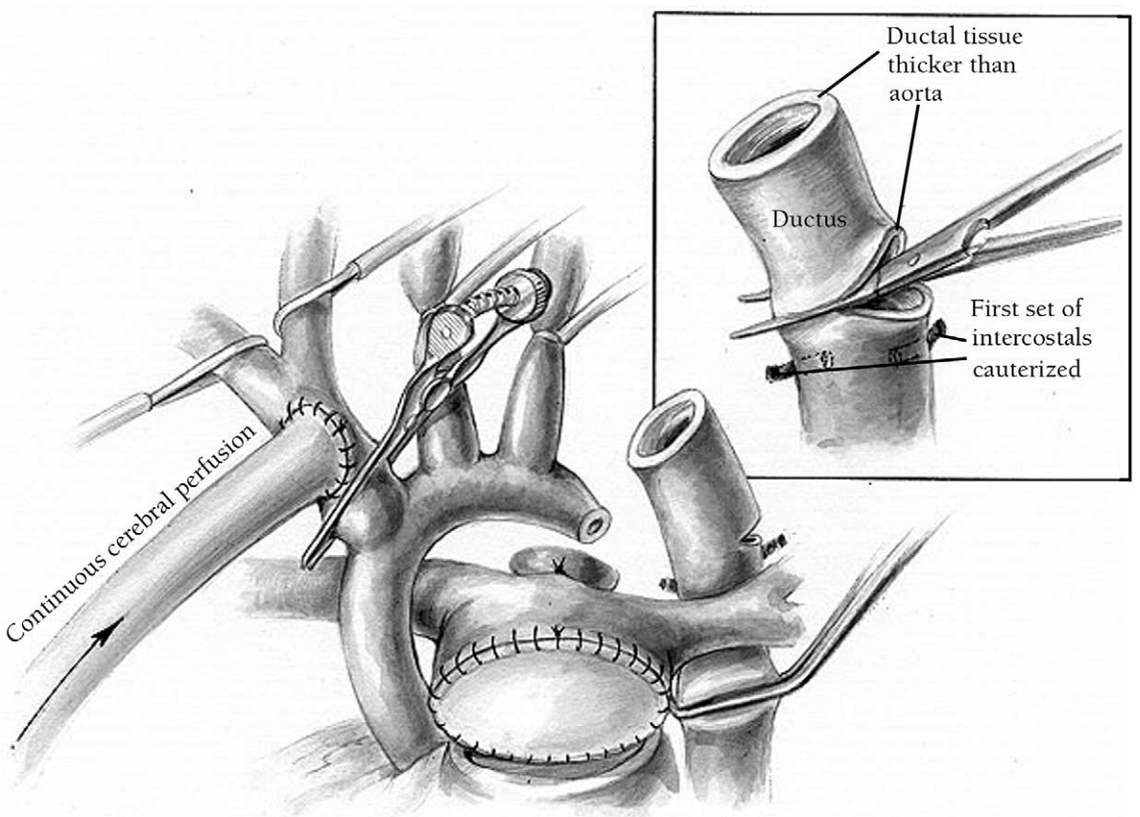

Figure 9 Initiation of continuous cerebral perfusion and excision of the ductus arteriosus. After delivering cardioplegia and completing the atrial septectomy, cerebral perfusion is initiated. A bulldog clamp or Cooley carotid clamp is placed across the origin of the innominate artery, the snares on the right subclavian artery and right common carotid artery are loosened, and antegrade flow is established. Because of the extensive collateral flow in a neonate, the snares on the left common carotid artery and left subclavian artery as well as the descending thoracic aortic clamp must remain on throughout the period of cerebral perfusion. The aortic isthmus is divided, and the ductus arteriosus is divided just distal to the ligature on the pulmonary artery side of the ductus. All residual ductal tissue is excised from the descending thoracic aorta. Two indicators are used to ensure that the ductal tissue is completely excised. First, the arterial duct is thicker walled and the texture more friable than the true descending aorta. Excision is continued until thin-walled, normal-textured artery is encountered. Second, the excision is continued to within 2 to 3 mm of the first set of intercostal branches; because the intercostals arise from the descending thoracic aorta and not the ductus arteriosus, this will further ensure complete excision of the ductal tissue.

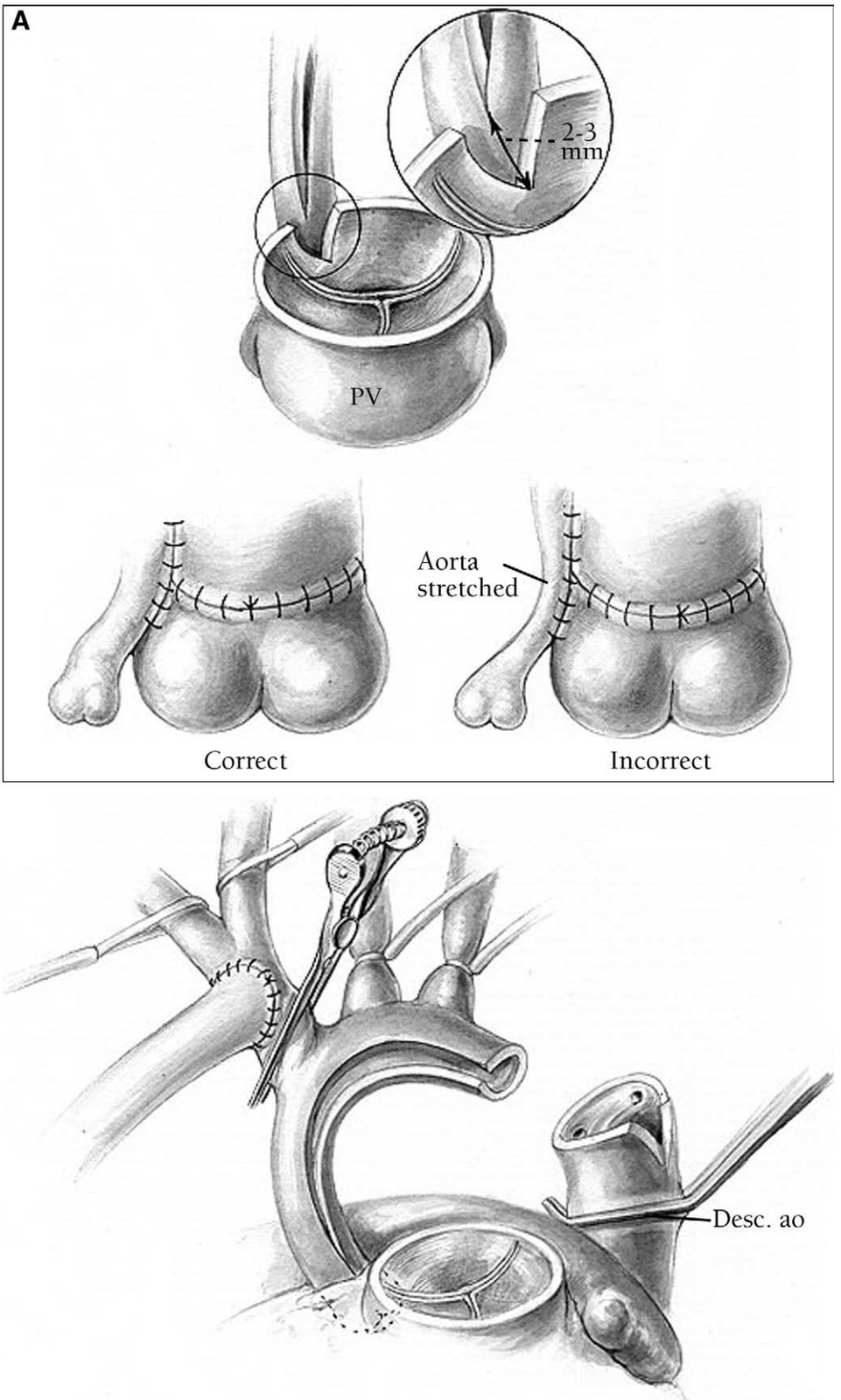

Figure 10 Arch reconstruction. The undersurface of the aortic arch is incised beginning at the open aortic isthmus and extending proximally down the ascending aorta (A). Great care must be taken with the diminutive ascending aorta to avoid spiraling down the ascending aorta. The incision must end precisely at the kissing point between the ascending aorta and the pulmonary root. A cutback is made in the pulmonary root at the site of implantation of the ascending aorta as shown in the top of the inset of Figure 10A. This cutback is in the posterior sinus of Valsalva just leftward of the commissure between the right and posterior sinus of Valsalva of the pulmonary valve. It is also important to end the aortic incision 2 to 3 mm cephalad to the level of the pulmonary root cutback.

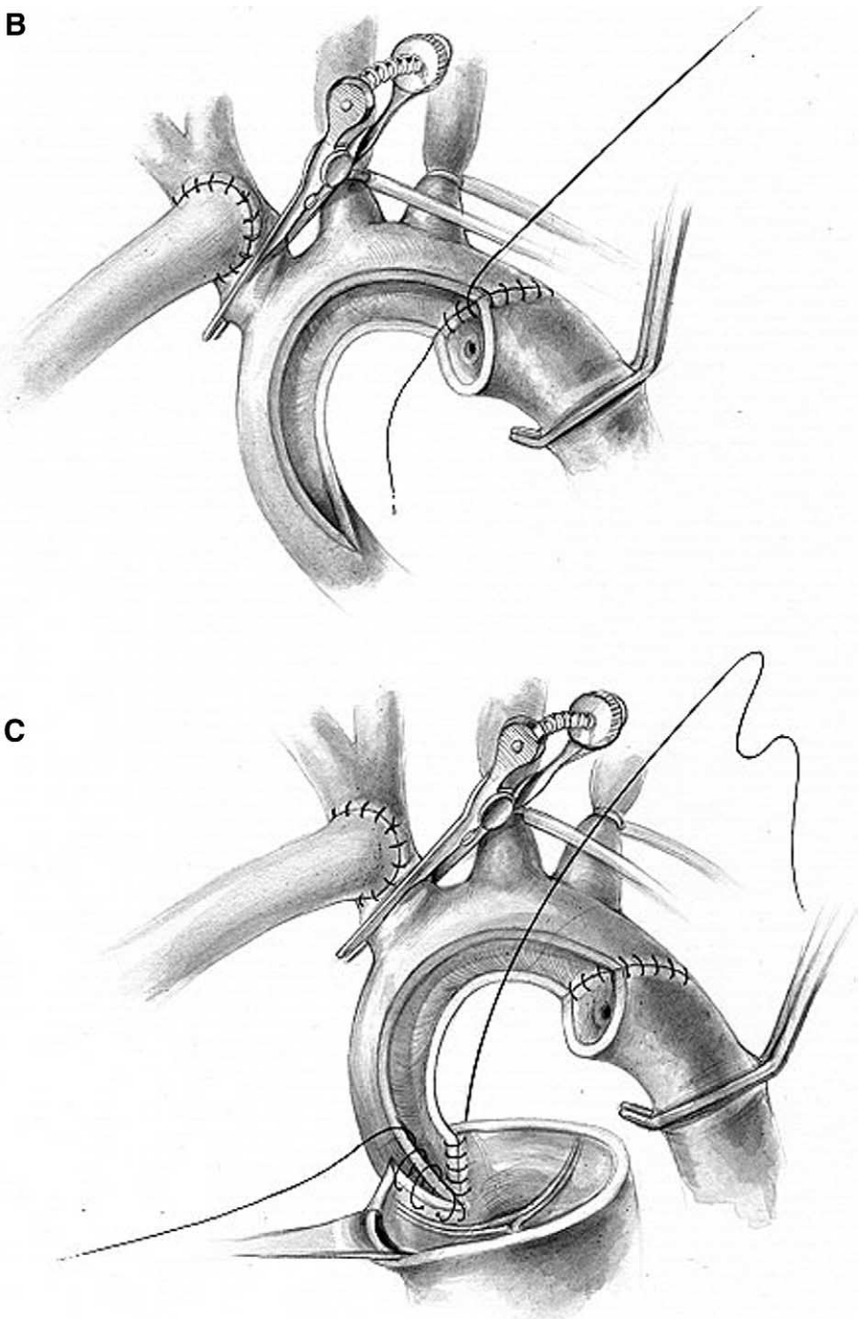

Figure 10 (Continued) This results in a small degree of redundancy in the proximal ascending aorta and ensures a patulous nonstenotic connection between the aortic root and the pulmonary root as shown in the lower left of the inset of Figure 10A. If the ascending aorta is incised too far proximally, a short segment of ascending aorta will be left, and stretching of this narrow caliber vessel (pictured in the lower right of the inset of Figure 10A) will result in stenosis and coronary insufficiency. Finally, a cutback is made into the posterior left lateral descending thoracic aorta. The proximity of the intercostal vessels to the proximal end of the descending thoracic aorta can be seen in Figure 10A and indicates the extent of ductal excision. Mobilization of proximal descending aorta is necessary to permit the reconstruction of the distal aortic arch. As shown in Figure 10B, the opened distal arch is interdigitated into the cutback in the proximal descending aorta. The clamp on the descending thoracic aorta assists with pulling it up and approximating it to the distal arch. Figure 10C shows the connection between the ascending aorta and aortic root, accomplished with a running suture technique.

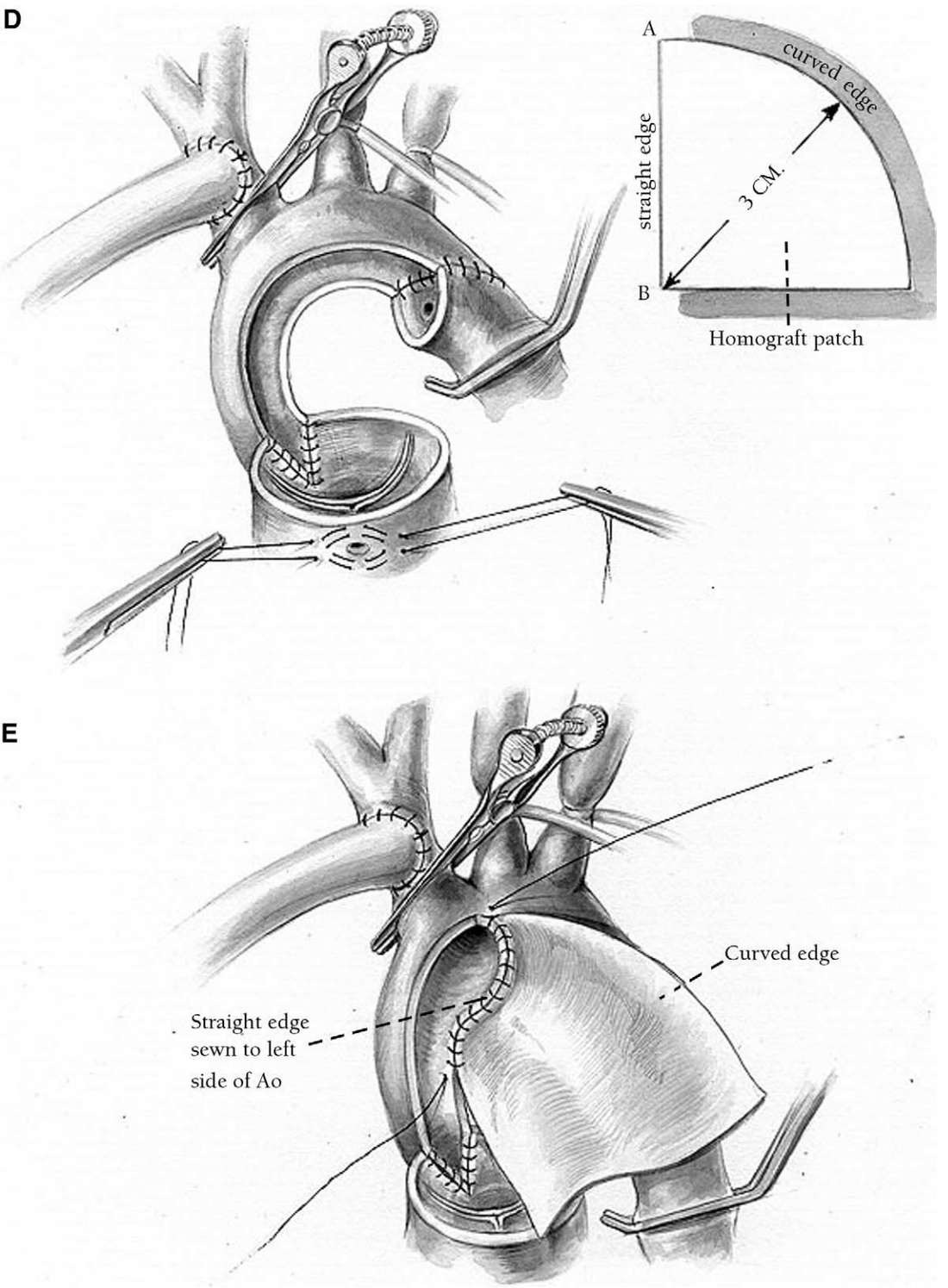

Figure 10 (Continued) While the pulmonary root is opened, the stab incision in the pursestring in the proximal pulmonary artery is made under direct vision to avoid injury to the pulmonary valve (Figure 10D). A pulmonary branch patch is used for completion of the arch reconstruction and creation of the neoascending aorta. This patch is a quarter circle with a radius of 3 cm. The straight edge of the circle from 12 o'clock to the center is sewn down the left edge of the ascending aorta (Figure 10E).

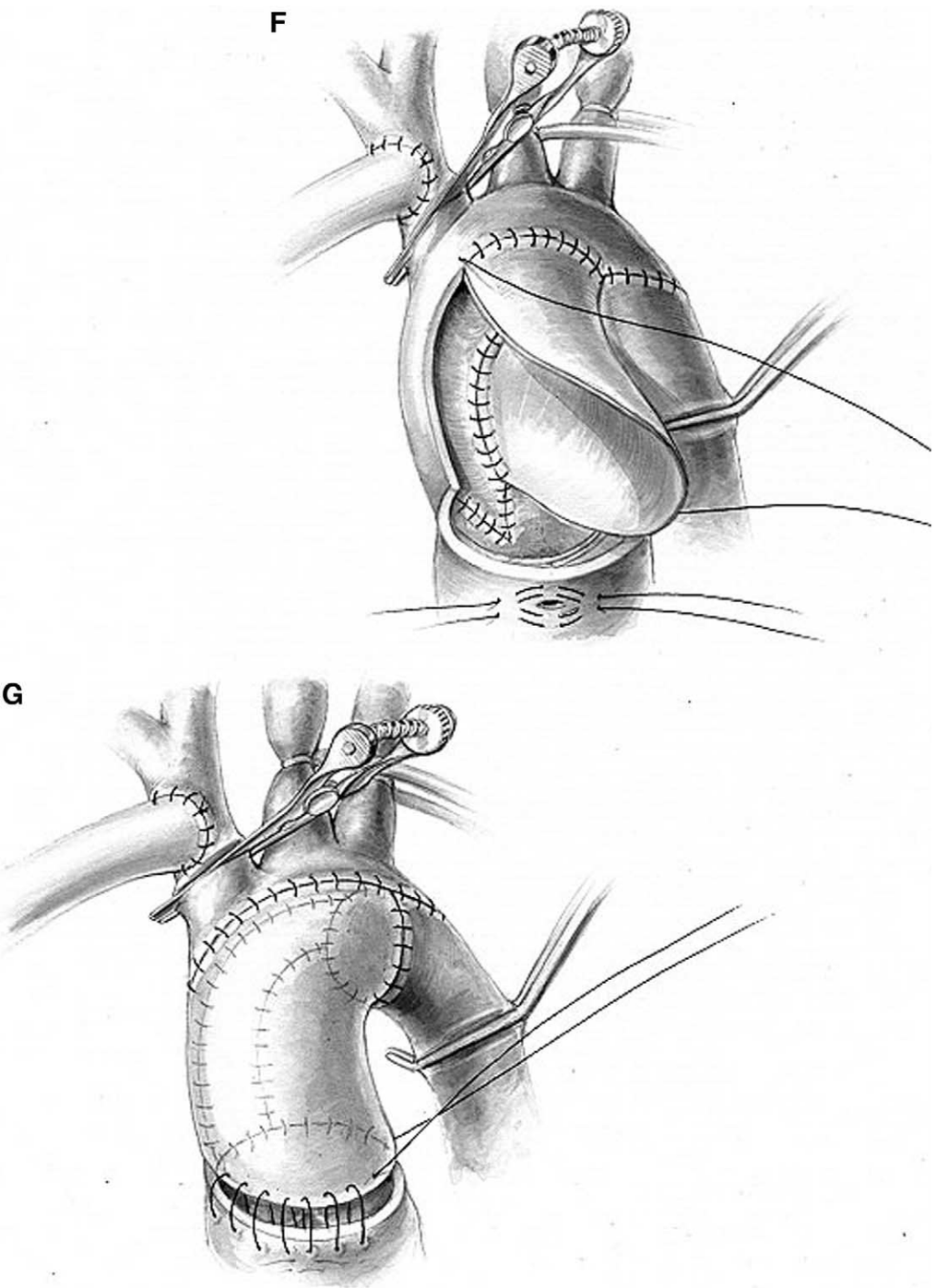

Figure 10 (Continued) The curved edge is sewn to the right edge of the ascending aorta (Figure 10F). When the junction of the ascending aorta and pulmonary root is reached, excess patch is trimmed, and the proximal connection between the patch and the pulmonary root is completed (Figure 10G).

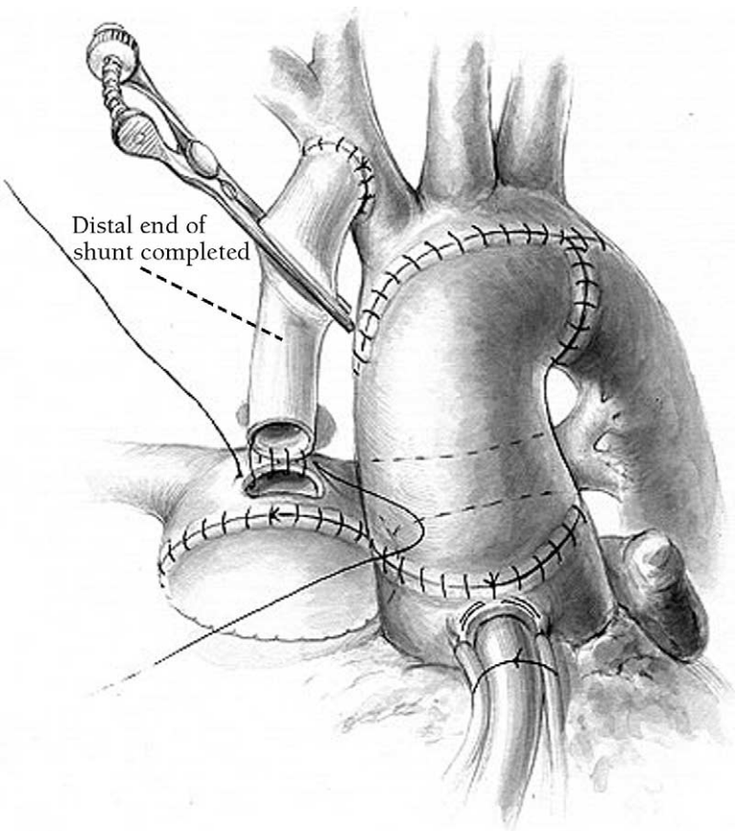

Figure 11 Completion of the distal innominate artery-to-pulmonary artery shunt. After reconstruction of the arch, ascending aorta, and completion of the Damus-Kaye-Stanzel anastomosis, the arterial cannula is transferred from the Gore-Tex graft to the neoaortic root. Air is evacuated from the neoaortic root and arch. This can be accomplished by tightening snares on the right common carotid artery and right subclavian arteries and giving some turns on the arterial cannula while evacuating any residual air through the Gore-Tex graft. After de-airing, the patient is placed in the Trendelenburg position, the clamp on the descending aorta is removed, cardiopulmonary bypass is resumed, snares on the brachiocephalic vessels are released, and rewarming is initiated. The central pulmonary confluence (distal main pulmonary artery) is moved to the right side of the ascending aorta. Early complete mobilization of the branch pulmonary arteries facilitates this maneuver. An arteriotomy is made in the central pulmonary artery confluence close to the patch suture line. The Gore-Tex graft is trimmed to a length of about 10 mm, and an anastomosis is constructed between the graft and the pulmonary artery. The central position of the shunt helps to promote equivalent growth of the branch pulmonary arteries.

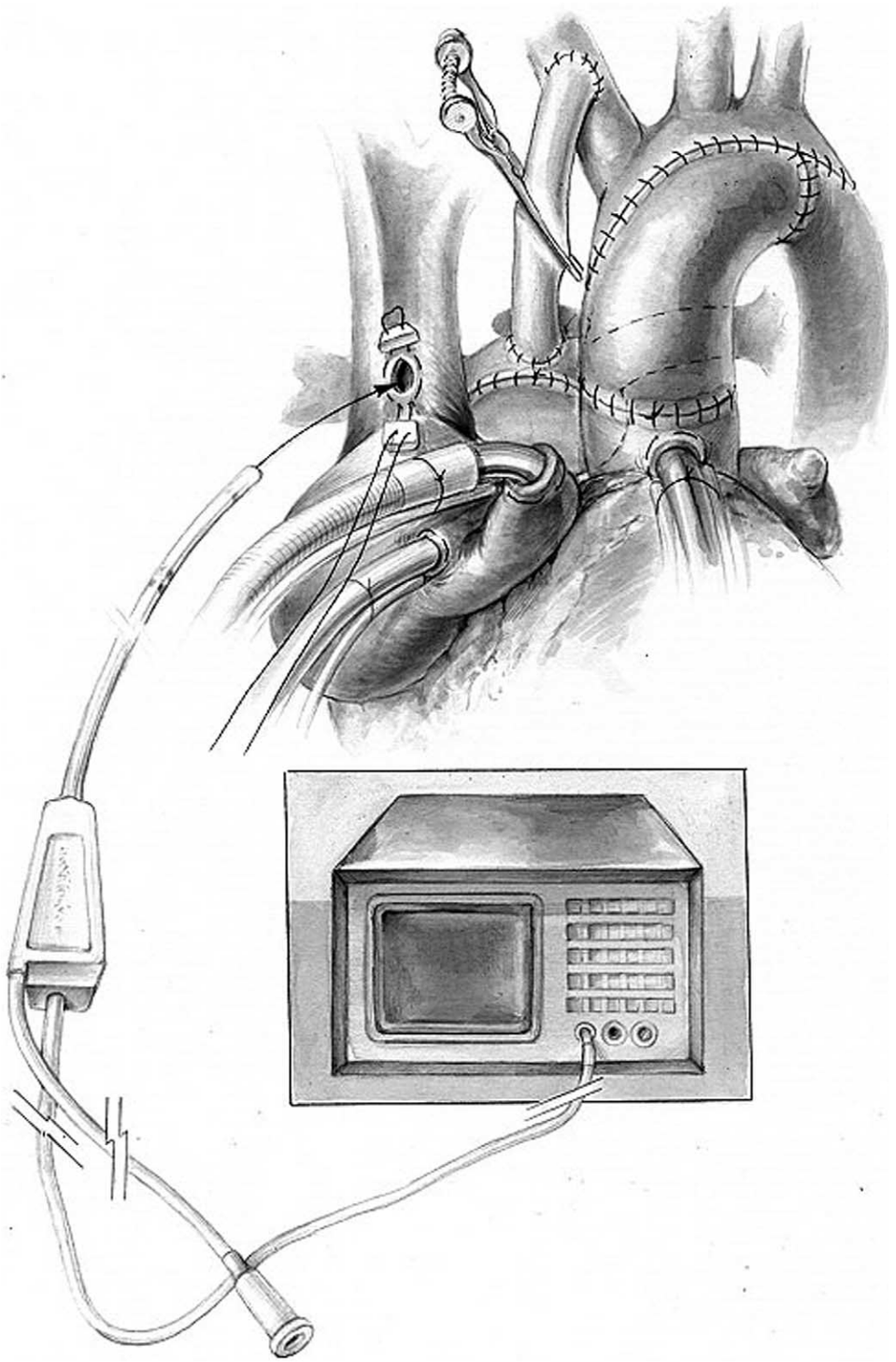

Figure 12 As rewarming is continued, the 4F oximetric catheter (Abbot Laboratories, North Chicago, IL) is placed through a small pledget-supported pursestring in the superior vena cava just cephalad to the superior vena cava right atrial junction. Only 1 to 2 mm of the catheter is positioned within the superior vena cava. No other lines are positioned within the superior vena cava to eliminate the risk of thrombosis. The pledgeted pursestring allows the line to be extracted in the intensive care unit several days after surgery.

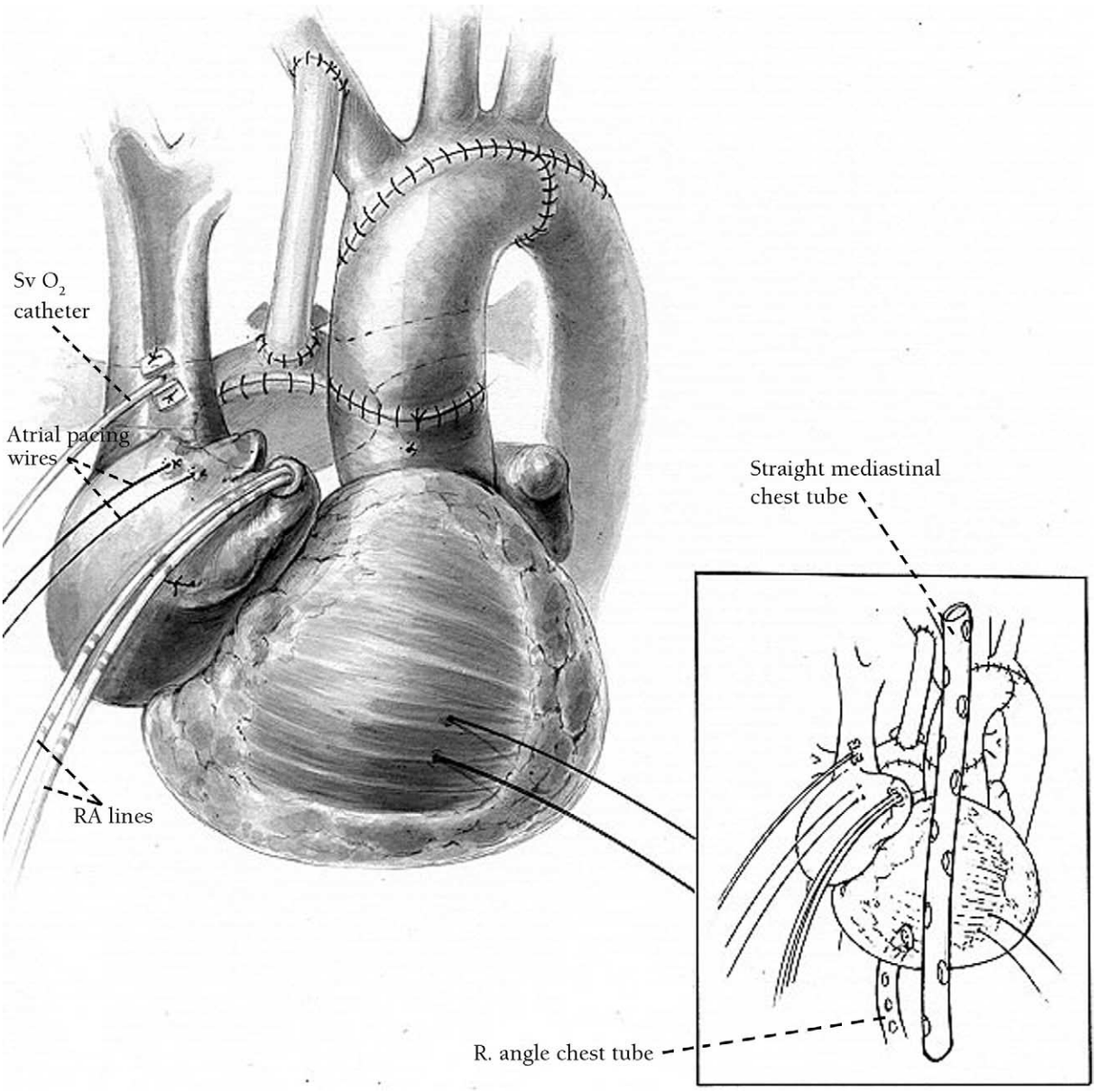

Figure 13 Completion of the operation. After rewarming to a bladder temperature greater than 35°C, inotropic support is initiated, the vent is removed, and mechanical ventilation is resumed. The heart is allowed to fill and ejection observed, and as a normal pulsatile pressure is achieved, the shunt is opened. If after several minutes hemodynamics remain satisfactory, modified ultrafiltration is initiated. After completion of modified ultrafiltration, the venous cannula is removed and protamine administered. When hemostasis is developing and the need for volume infusion is minimal, the arterial cannula is removed. Commonly, the sternum is left open. A silastic patch is used to close the mediastinum and secured to the skin edge with adhesive. Transparent dressings (Tegaderm, St. Paul, MN) are used to further secure the silastic patch to the skin. The skin edges remain healthy, and routine subcuticular skin closure can be accomplished even several days after the original operation. Should hemodynamics prove unsatisfactory after weaning from bypass, residual lesions must be sought out and corrected. It is rare to have residual arch obstruction or a restrictive atrial septal defect, and therefore most problems are related to overall ventricular function or problems with the systemic-to-pulmonary artery shunt. We generally aim for an SaO_2 of at least 75%, and in the face of unacceptably low SaO_2, upsizing of the shunt should be considered. Arterial saturations greater than 90% are tolerable, unless there is evidence of decreased systemic perfusion as indicated by low SvO_2 or a diastolic pressure of less than 30 mm Hg, in which case downsizing of the shunt may be necessary.

Comments

The operative technique depicted in the figures above has evolved over time, and innovations described by other surgeons have been adopted to improve results, increase efficiency of the procedure, and/or limit the duration of total support. We felt it was prudent to limit the duration of deep hypothermic circulatory arrest by using continuous cerebral perfusion as described by Pigula and colleagues.[7] Division of main pulmonary artery and subsequent patching of the pulmonary artery confluence are accomplished during cooling as described by Fraser and Mee.[8] We perform a coarctectomy and interdigitate the distal arch into the proximal descending thoracic aorta, with an aim to eliminate any residual ductal tissue and maximize the native tissue-to-native tissue connection of the arch reconstruction. This approach was borrowed from Van Arsdell and colleagues (personal communication, September 2002) and has virtually eliminated recurrent coarctation in our experience. Routinely, a cutback is made into the pulmonary root leftward of the pulmonary valve commissure that is adjacent to the ascending aorta. The incision in the ascending aorta is ended a few millimeters above the cutback in the pulmonary root to ensure there is no stretching of the ascending aorta. These 2 steps increase the size of the connection between the ascending aorta and the pulmonary artery, create mild redundancy in the ascending aorta-pulmonary root connection preventing stretch, and finally, displace this connection away from the tripartite connection of the pulmonary root ascending aorta and homograft patch, decreasing the potential for twisting or distortion. These steps minimize the potential for coronary ischemia.

Postoperative management, which has been detailed in previous publications, centers on maintaining adequate systemic oxygen delivery with continuous monitoring of the superior vena cava saturation as an approximation of venous oxygen saturation (SvO_2). We target an SvO_2 greater than 50%. Other hemodynamic parameters are adjusted to meet this goal. An atrioventricular (AV) sequential rhythm is essential. Although postoperative heart block is exceedingly rare in this group of patients, occasionally we have found that AV pacing with a shortened PR interval results in improved cardiac output and less tricuspid valve insufficiency among patients with an abnormal tricuspid valve anatomy. Mean arterial pressure is maintained at or above 45 mm Hg and diastolic blood pressure equal to or greater than 30 mm Hg. Preload is titrated to meet the goals of venous saturation and blood pressure and, commonly, the central venous pressure is between 6 and 12 mm Hg. We aim to have an arterial saturation of at least 75%, but we do not have a strict upper limit of arterial saturation, and therefore the fractional concentration of oxygen in inspired gas is not limited. The fractional concentration of oxygen in inspired gas is typically maintained no lower than 40% for the first 24 hours to prevent pulmonary venous desaturation. The hematocrit is adjusted as dictated by the SvO_2 and is generally maintained above 45%. Two-site near-infrared spectroscopy is used in the early postoperative period to minimize the potential for decreased cerebral oxygen delivery, which is easily identified and managed by maintaining satisfactory cardiac output along with mild hypercapnea (partial pressure of carbon dioxide >45 mm Hg).[5] The sternum is commonly but not routinely left open after the operation, and delayed sternal closure is performed most often on postoperative day 1 or 2. After extubation, the hospital course centers on achieving adequate oral intake. We target 120 kCal/kg/d with a goal to demonstrate consistent weight gain as one of the discharge criteria. If adequate oral feeding cannot be achieved, a gastrostomy tube is surgically placed. Patients are not discharged with nasogastric tube feeds to minimize the risk of respiratory complications that can be a source of interstage death. Patients are discharged with supplemental oxygen if saturations are not maintained above 75% on room air. All patients are placed in the home monitoring program at discharge.[9]

Among 116 patients with true HLHS as defined by Ashburn and colleagues,[10] who underwent stage I palliation at the Children's Hospital of Wisconsin between July of 1996 and December of 2004, there have been 6 early deaths (within 30 days of operation or during the primary hospitalization), for an early survival rate of 94.8%. We have used continuous cerebral perfusion in 67 of these 116 patients, with an early survival of 97% (65/67). Survival for patients undergoing stage I palliation with continuous cerebral perfusion is 93% ± 3% at 1 year and 90% ± 4.5% at 3 years. It is noteworthy, however, that this improvement in intermediate survival corresponds to the implementation of our home monitoring program and cannot be entirely attributed to the change in our perfusion strategy.

Although strategies for stage I palliation of HLHS continue to evolve, common to all strategies are the goals of achieving unobstructed systemic flow, including unimpeded coronary blood flow, a nonrestrictive atrial septal defect, and appropriate limitation of pulmonary blood flow. The surgical procedure outlined above is a tested, reliable, and reproducible way to achieve the goals of stage I palliation and has yielded excellent results. Whether any of the modifications of stage I palliation of HLHS prove to be superior is as yet undetermined. Although a carefully performed operation is an essential first step, in this fragile group of patients, data from our program would indicate that sustained quality survival is also the product of ongoing objective assessment of the patients' circulatory status.

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