Operative Techniques in Thoracic and Cardiovascular Surgery

Stage I—The Philadelphia Approach

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The term hypoplastic left heart syndrome (HLHS) describes a group of cardiac malformations that result in a diminutive or absent left ventricle and a hypoplastic ascending aorta; these structures are inadequate to supply the systemic circulation. Mitral or aortic valve hypoplasia or atresia may accompany the syndrome. The circulation is dependent on the patency of the ductus arteriosus as well as on mixing of systemic and pulmonary venous blood.[1] This syndrome is the most common severe congenital heart defect; it represents 7 to 9% of all congenital cardiac anomalies.[2]

The earliest attempt to provide palliation for HLHS involved a graft that connected the right ventricle to the descending aorta, and pulmonary blood flow was controlled with bilateral banding. These attempts were troubled by pulmonary arterial distortion, but they laid the groundwork for the development, largely by Dr. William I. Norwood, of the first successful newborn palliative reconstruction. The current staged approach to palliation described by Norwood[3] has changed little in the past two decades, but there are some recently proposed, potentially important technical modifications currently under investigation. An alternative therapy is cardiac transplantation; this strategy was pioneered by Bailey and colleagues, but it is limited by donor organ availability.[4]

The principles of the staged reconstruction are similar to those for other single ventricle anomalies. The goals of the first stage procedure are (i) creation of an unobstructed connection between the aorta and the systemic ventricle, (ii) regulation of pulmonary blood flow, and (iii) creation of an unobstructed interatrial communication.[3,5] Retrograde perfusion of the ascending aorta must be provided to avoid coronary underperfusion. Significant AV valve regurgitation may be addressed; alternatively very severe regurgitation may contraindicate a staged approach in favor of primary transplantation.

The second stage is performed at 3 to 6 months of age and involves division of the systemic to pulmonary shunt and creation of a bidirectional Glenn or hemi-Fontan connection. This serves to unload significantly the single ventricle and also to create a more reliable source of pulmonary blood flow than the previously shunt-dependent circulation. The final stage is a modified Fontan–Kreutzer procedure in which total cavopulmonary connection is achieved with an intraatrial lateral tunnel or extracardiac connection.[3]

Preoperative Preparation and Anesthetic Considerations

Previously undiagnosed HLHS manifests as severely compromised systemic perfusion when the ductus arteriosus begins to close. Significant organ dysfunction and frank shock can ensue. Echocardiography is the mainstay of diagnosis and therefore must be undertaken rapidly when HLHS is suggested. The clinical situation is greatly worsened by the presence of a restrictive atrial septal defect (2 to 5% of cases), which causes an obstruction of pulmonary venous outflow with resultant congestion, hypoxemia, and pulmonary disease. Restrictive atrial septal defects require dilation in the catheterization laboratory or emergent septectomy or complete surgical correction.

Medical therapy for HLHS is undertaken as soon as the diagnosis is made and involves maintaining ductal patency with a continuous infusion of prostaglandin E1 (0.025 to 0.05 μg/kg/min). Spontaneous ventilation is preferred in these patients, although many will develop apnea related to the prostaglandins and require mechanical ventilation. Excessive oxygen administration is avoided in an attempt to balance pulmonary and systemic flow. Occasionally inspired carbon dioxide is helpful in controlling pulmonary overcirculation, and it may improve cerebral circulation. It is generally possible to stabilize these patients medically and to allow for recovery of end-organ dysfunction before surgical intervention.

Operative Technique at the Children's Hospital of Philadelphia

The operative technique for the Stage I reconstruction for HLHS utilized at the Children's Hospital of Philadelphia relies heavily on the technique described by Dr. Norwood more than 20 years ago.[3,5] Deep hypothermic circulatory arrest is employed for the reconstruction. We do not employ retrograde or selective cerebral perfusion, although these techniques are favored by some centers. We do not believe that the data available to date show any clear benefit arising from

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selective or retrograde perfusion as compared with circulatory arrest[6-9]; moreover these perfusion techniques can be cumbersome and interfere with precise reconstruction.

Pulmonary blood flow is generally provided via a modified Blalock–Taussig shunt from the right innominate artery to the pulmonary artery. This shunt is relatively small, sized 3.5 to 4.0 mm, and is chosen so as to be the limiting factor for pulmonary blood flow. Providing limitation of pulmonary blood flow at the level of the shunt facilitates postoperative management by rendering traditional manipulations of the ventilator to modify pulmonary vascular resistance unnecessary.[10] Recently a right ventricle to pulmonary artery conduit[11] has been employed in a minority of cases. The precise determination of the relative benefit of this conduit awaits the results of ongoing investigation.

The creation of the neoaorta begins with a side-to-side connection of the aorta to the pulmonary artery. The arch is augmented with a patch of pulmonary homograft, and resection of the coarctation segment is rarely performed. We believe that the tailoring of the patch is key to minimizing the risk of arch obstruction, and the details of our technique are described.

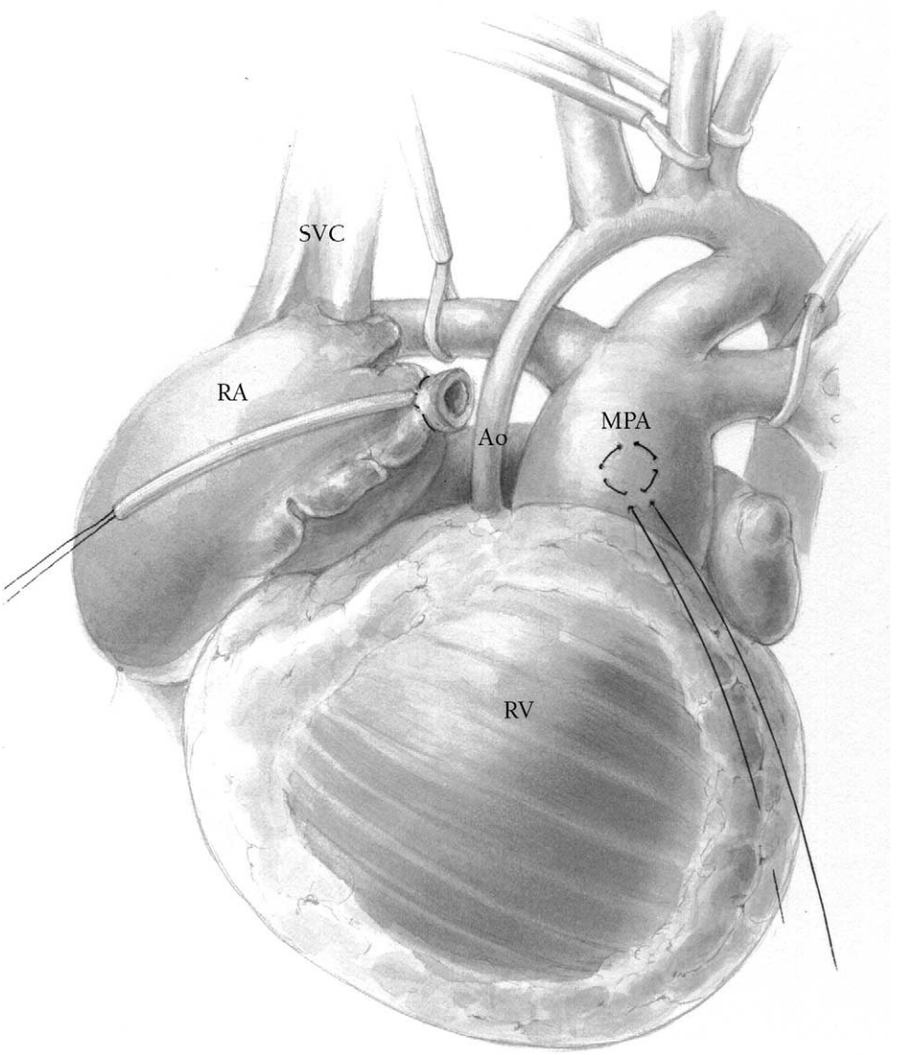

Figure 1 Initial exposure is via a median sternotomy. The thymus is removed and the pericardium opened in the midline. Lymphoid tissue between the superior vena cava and aorta overlying the trachea is removed to allow the shunt to lie properly. The diminutive aorta is fully mobilized off the pulmonary artery. The arch is mobilized into the descending aorta, beyond the ductal insertion. Tourniquets are passed around the arch vessels (except the innominate artery), and the right and left pulmonary arteries. Ao = aorta; MPA = main pulmonary artery; RA = right atrium; RV = right ventricle; SVC = superior vena cava artery.

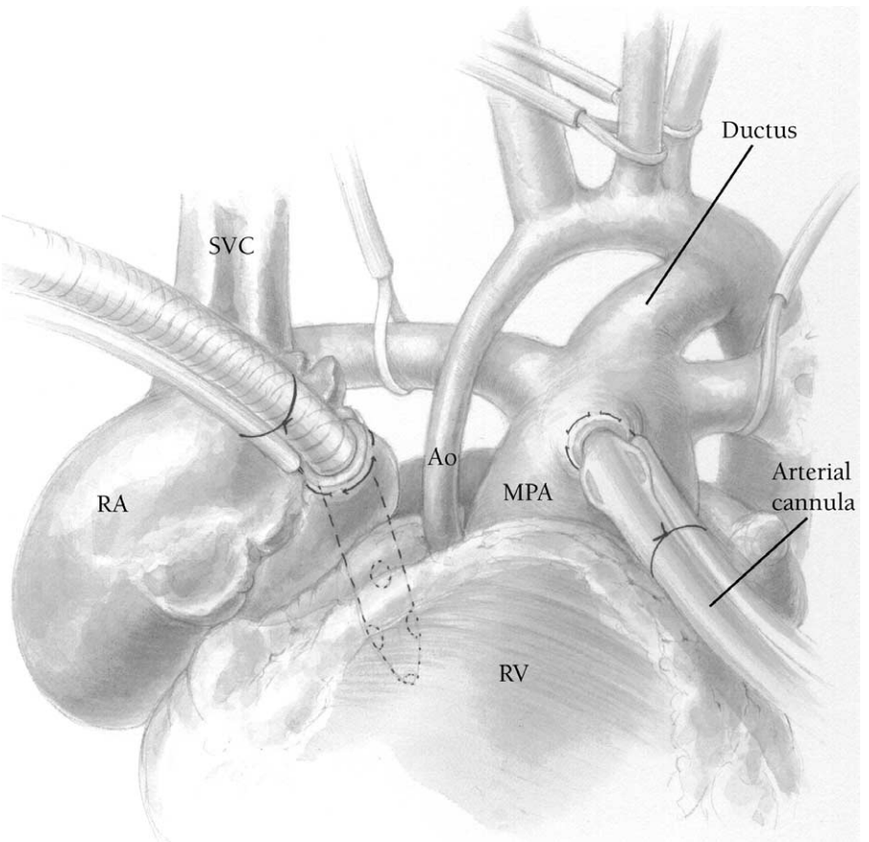

Figure 2 After systemic heparinization the pulmonary artery is cannulated proximal to its bifurcation, above the pulmonary valve sinuses, and a single venous cannula is placed in the right atrium. When cardiopulmonary bypass is instituted, the snares on the pulmonary arteries are tightened to direct perfusion through the ductus arteriosis. Ao = aorta; MPA = main pulmonary artery; RA = right atrium; RV = right ventricle; SVC = superior vena cava artery.

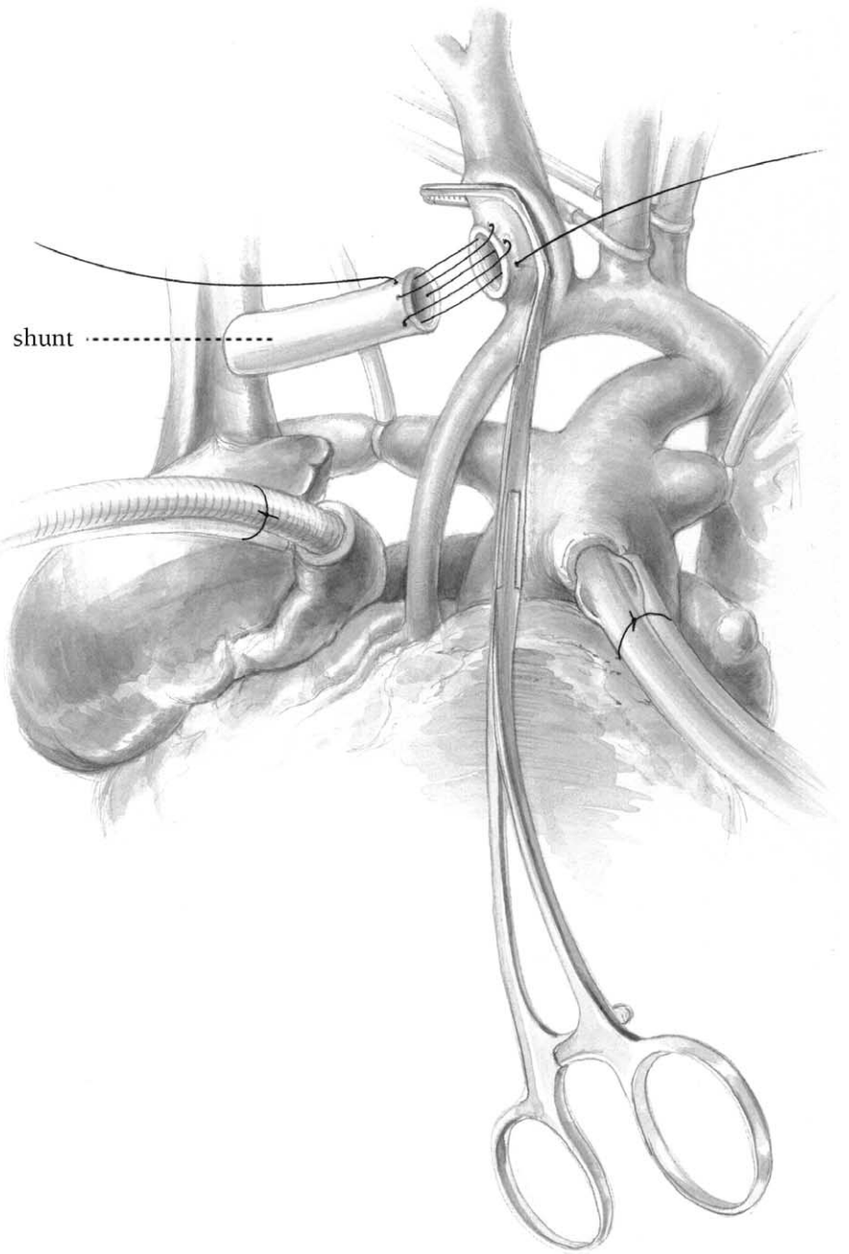

Figure 3 Bypass is instituted and the patient is cooled to 18°C. During cooling the proximal anastamosis of the shunt (usually 3.5 or 4.0 mm) is created to the proximal innominate artery with 7.0 monofilament suture. It is best to cut the shunt to its appropriate length before circulatory arrest, while the structures are still filled with blood. After completion of the shunt, it is gently occluded with a hemoclip. A tourniquet is then placed around the innominate artery.

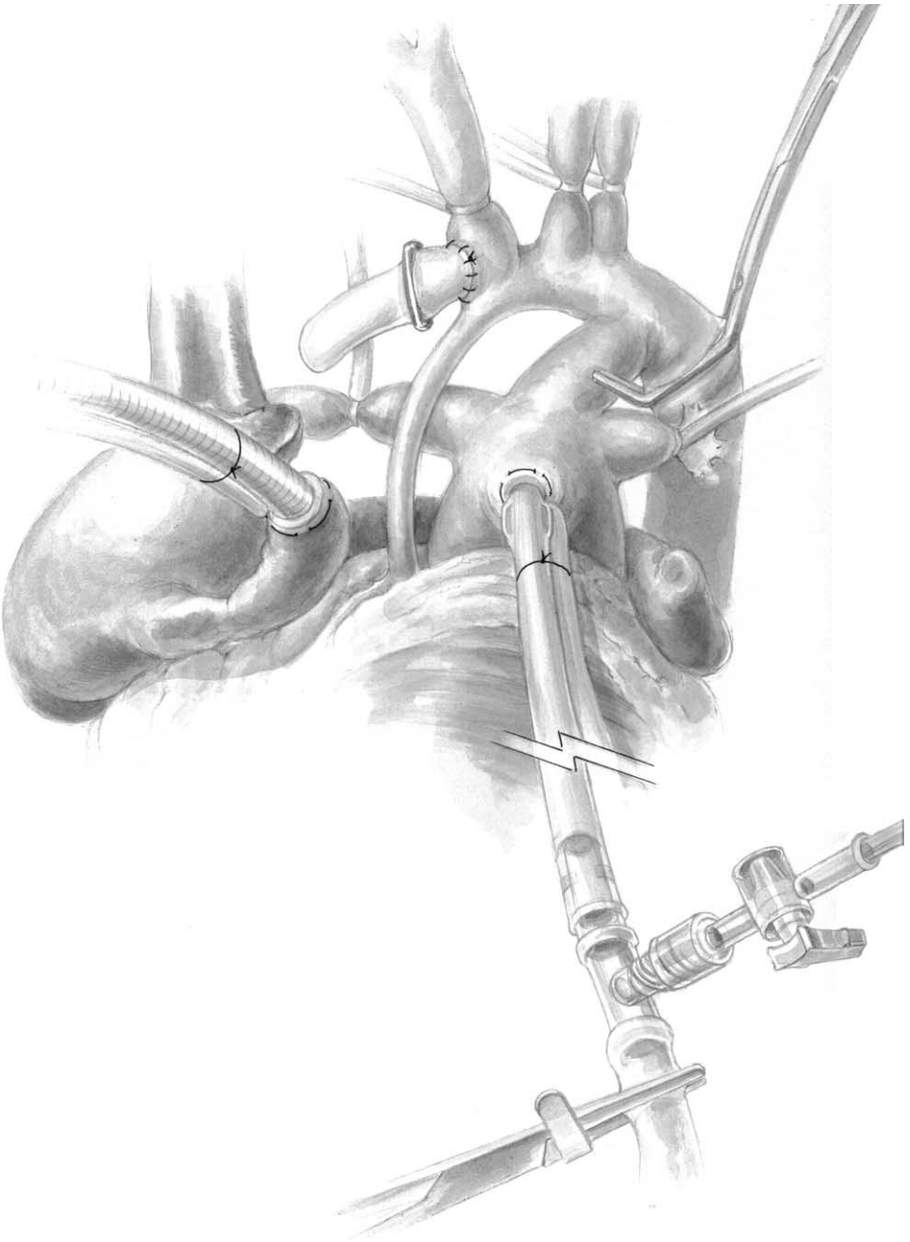

Figure 4 When the patient is adequately cooled, circulatory arrest is initiated. The snares on the aortic arch vessels and on the pulmonary arteries are tightened. With a clamp placed on the aorta distal to the ductal insertion, cardioplegia is administered via a sidearm connector on the arterial line. After administration of cardioplegia, the snares on the pulmonary arteries are removed.

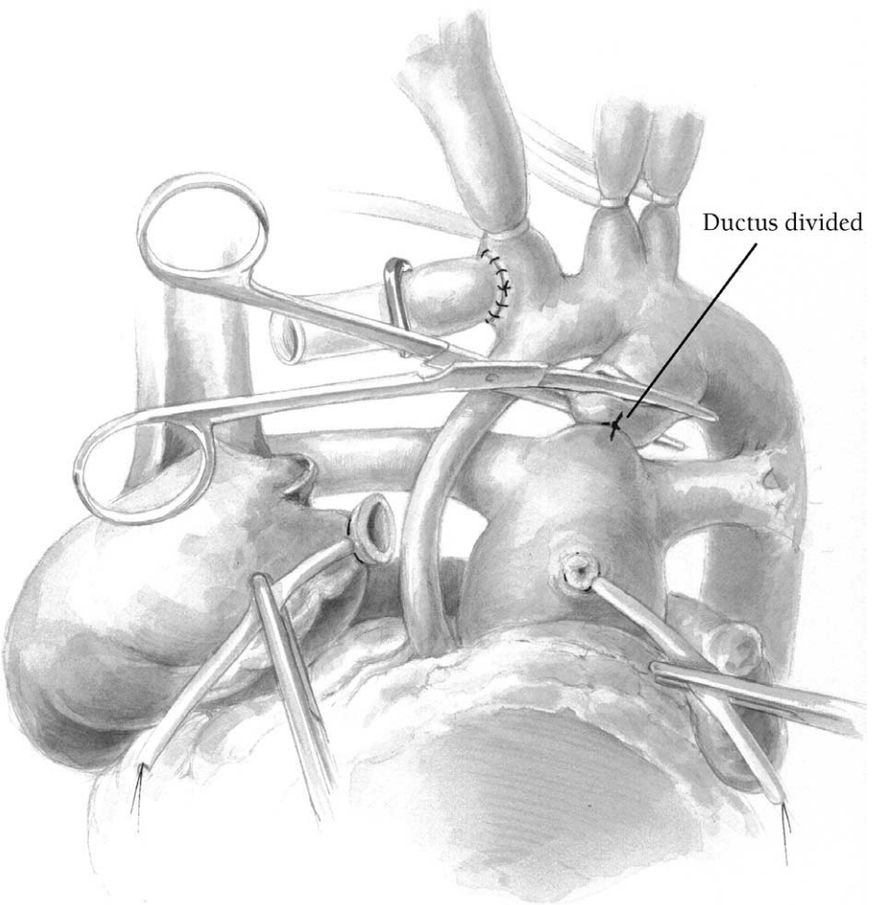

Figure 5 The ductus is tied and divided, leaving the tie on the pulmonary artery side.

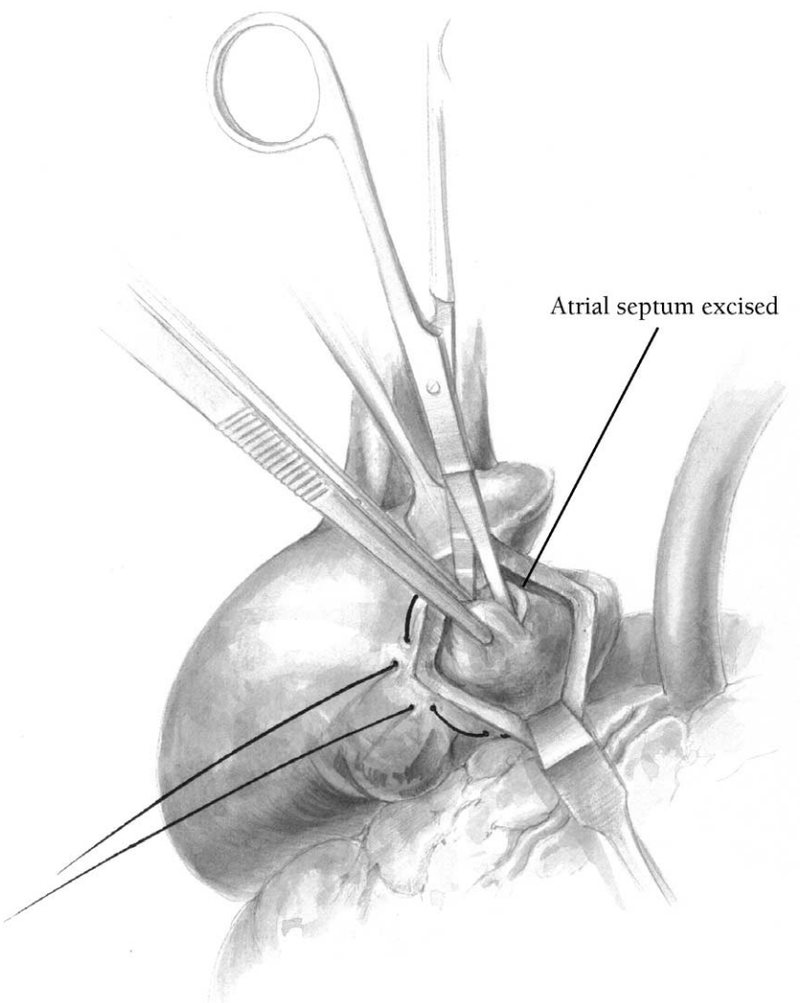

Figure 6 The atrial septum is excised through the venous purse-string or through a separate atriotomy.

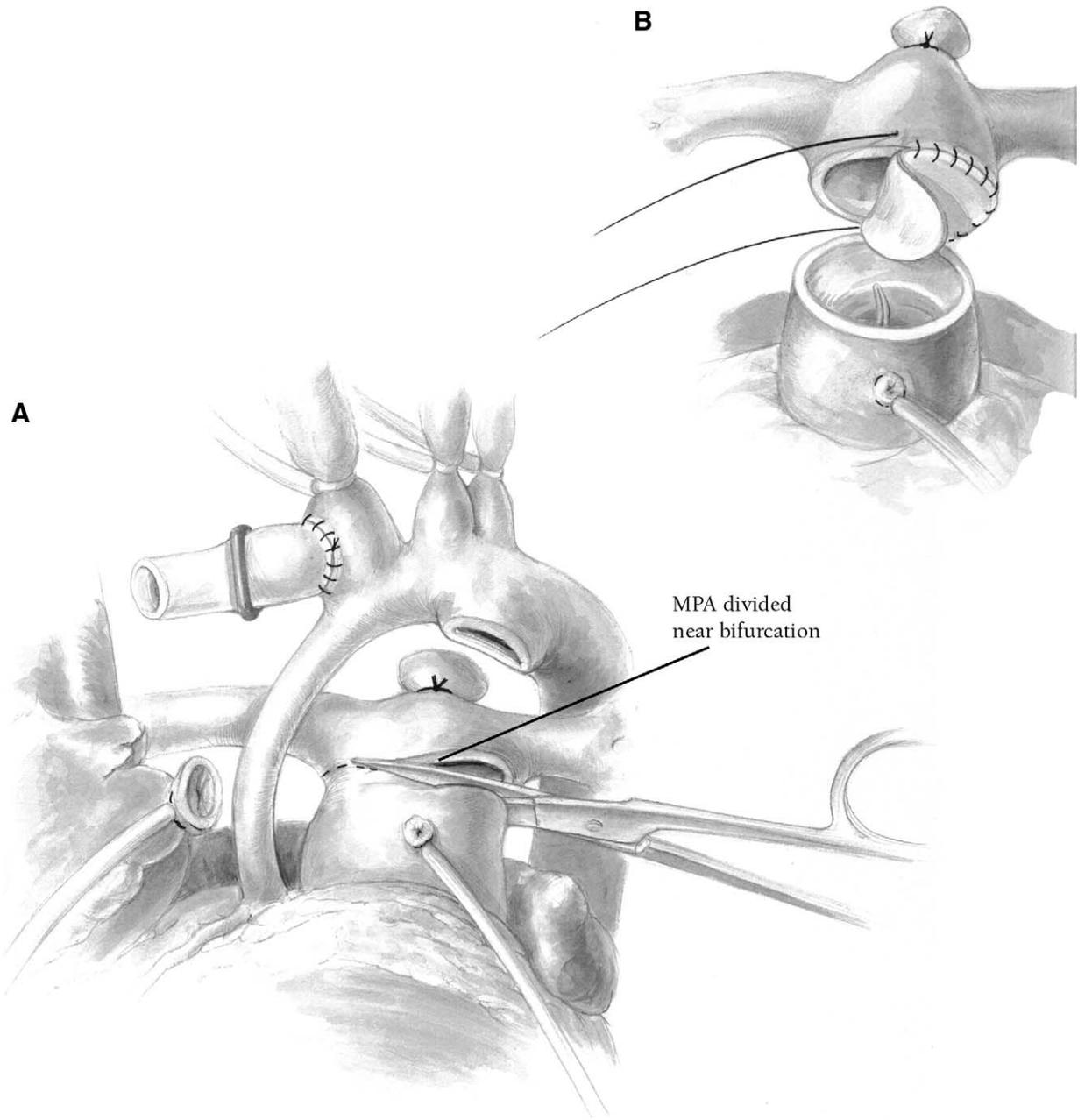

Figure 7 The pulmonary artery is divided transversely at the origin of the right pulmonary artery. Making this division at the origin of the right pulmonary artery ensures that the connection of the PA to the aorta is distal enough that it does not interfere with the coronary arteries. The distal pulmonary artery is closed primarily or with a pulmonary homograft patch. MPA = main pulmonary artery.

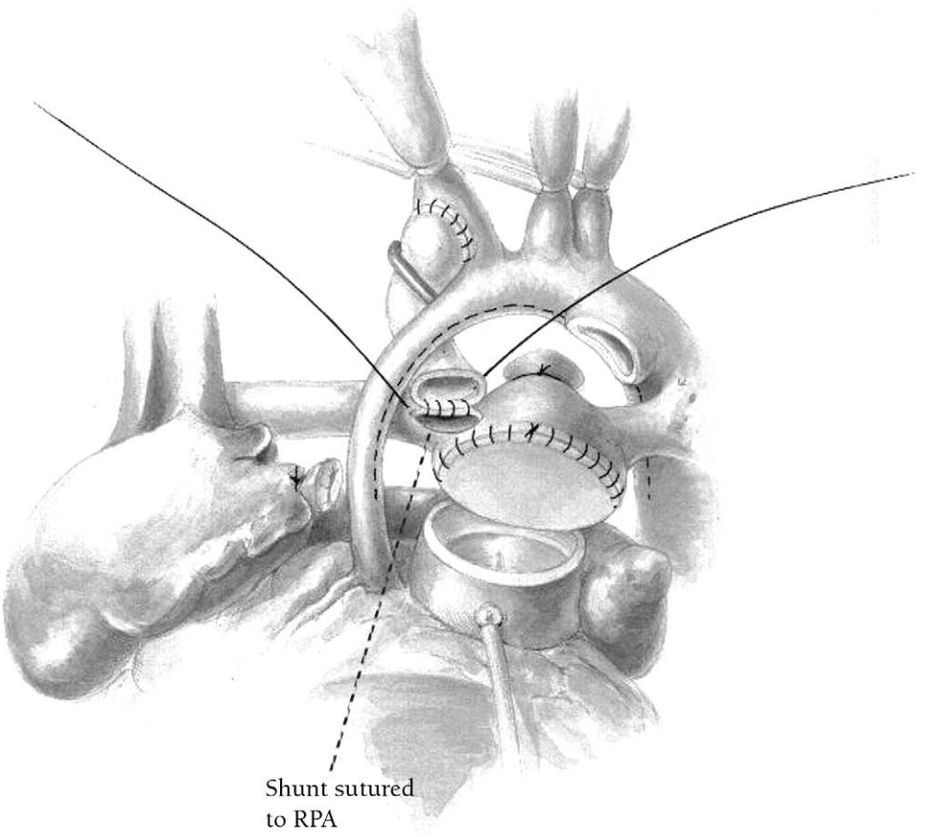

Figure 8 The distal anastomosis of the shunt is created to the right pulmonary artery very proximally (almost at the origin of the right pulmonary artery) with 7.0 monofilament suture. The hatched line demonstrates the incision to be made starting in the diminutive ascending aorta and carried beyond the ductal insertion. RPA = right pulmonary artery.

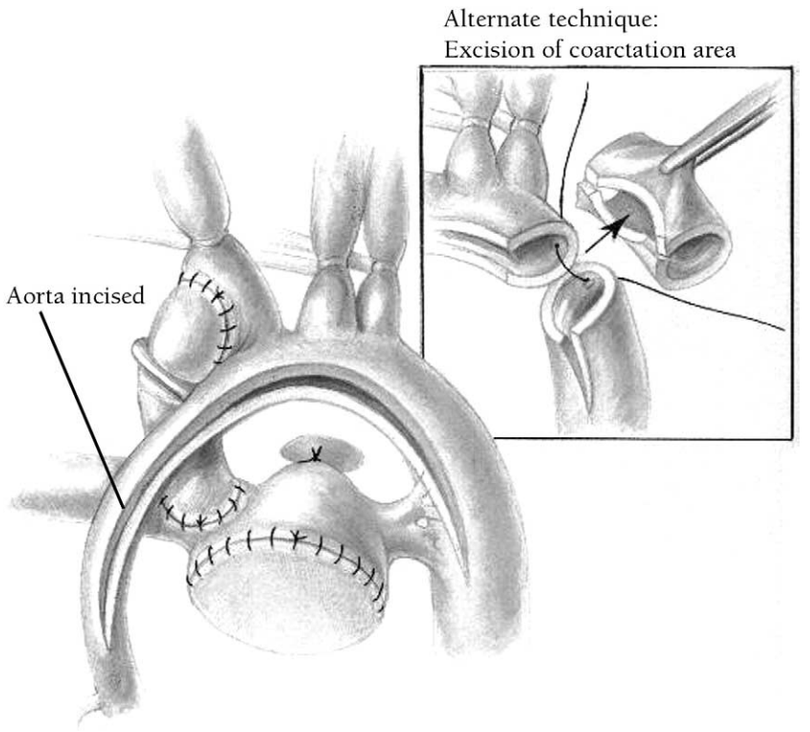

Figure 9 After the aorta is opened, the ridge of ductal tissue at the coarctation is excised. In a small proportion of cases the coarctation segment is resected (inset).

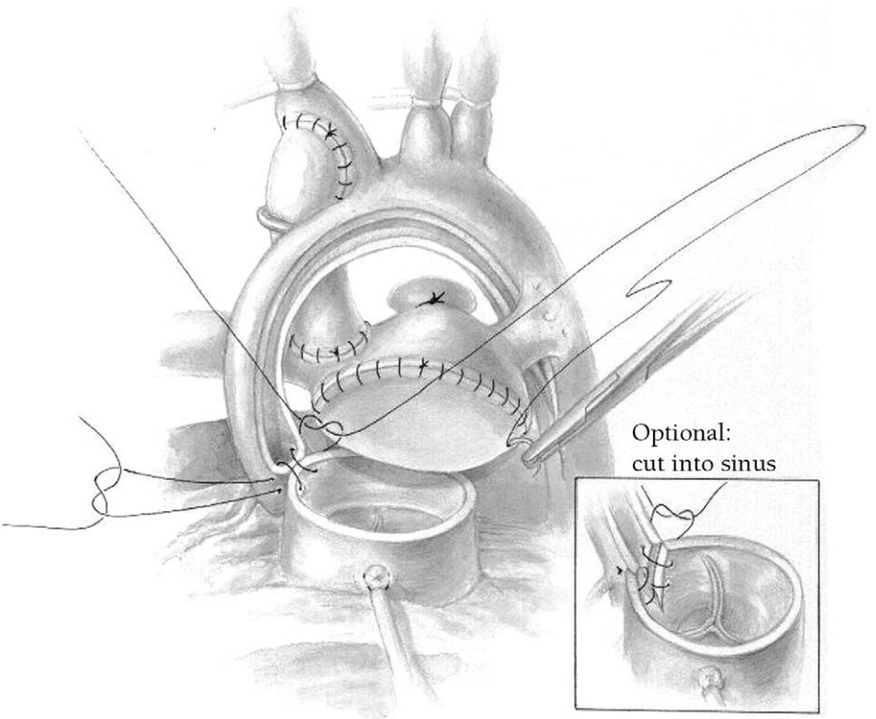

Figure 10 The creation of the neoaorta begins with approximation of the aorta and proximal pulmonary artery with several interrupted sutures of 7.0 prolene. Occasionally a "cutback" is created into the sinus of the pulmonary artery (inset), but usually the connection is created without this slit. The purpose of the "cutback" is to prevent limitation of coronary blood flow due to distortion of the diminutive aorta. If the "cutback" is made, the commissures of the pulmonary valve are generally positioned at the level of the aortic incision.

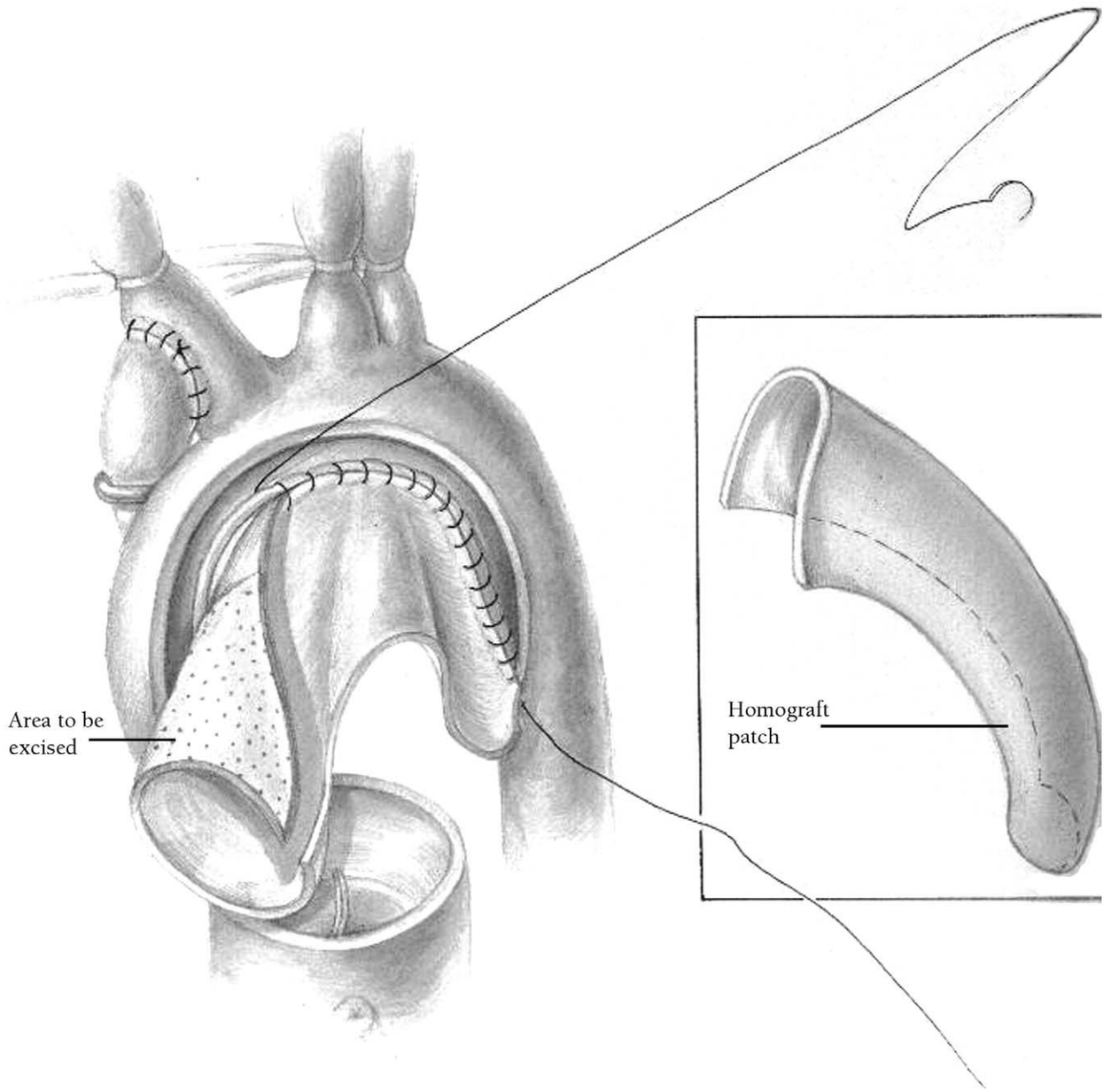

Figure 11A Next the aorta is augmented with a patch of pulmonary homograft, starting distally. The tailoring of the patch geometry is complex. Generally as the suture line of the patch is brought under the arch vessels, it becomes necessary to narrow the patch posteriorly by removing a piece of homograft (shown with hatchmarks), to avoid kinking and twisting of the reconstruction. In general it is better for the patch to be slightly too narrow rather than slightly too wide.

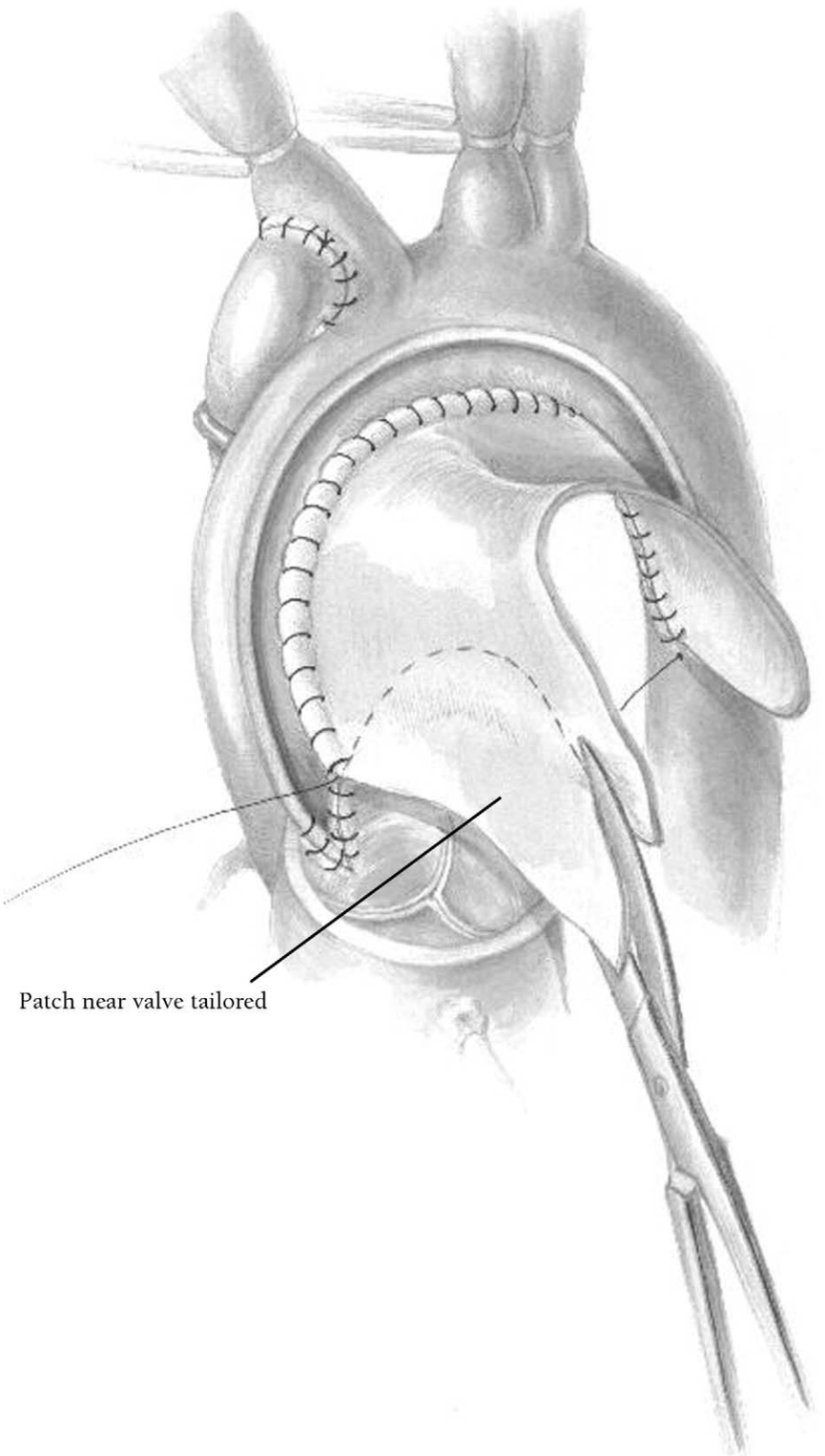

Figure 11B The portion of the patch that gets sewn to the pulmonary artery needs to be further tailored so as to be concave, by removing the hatched segment, also to avoid kinking of the reconstruction.

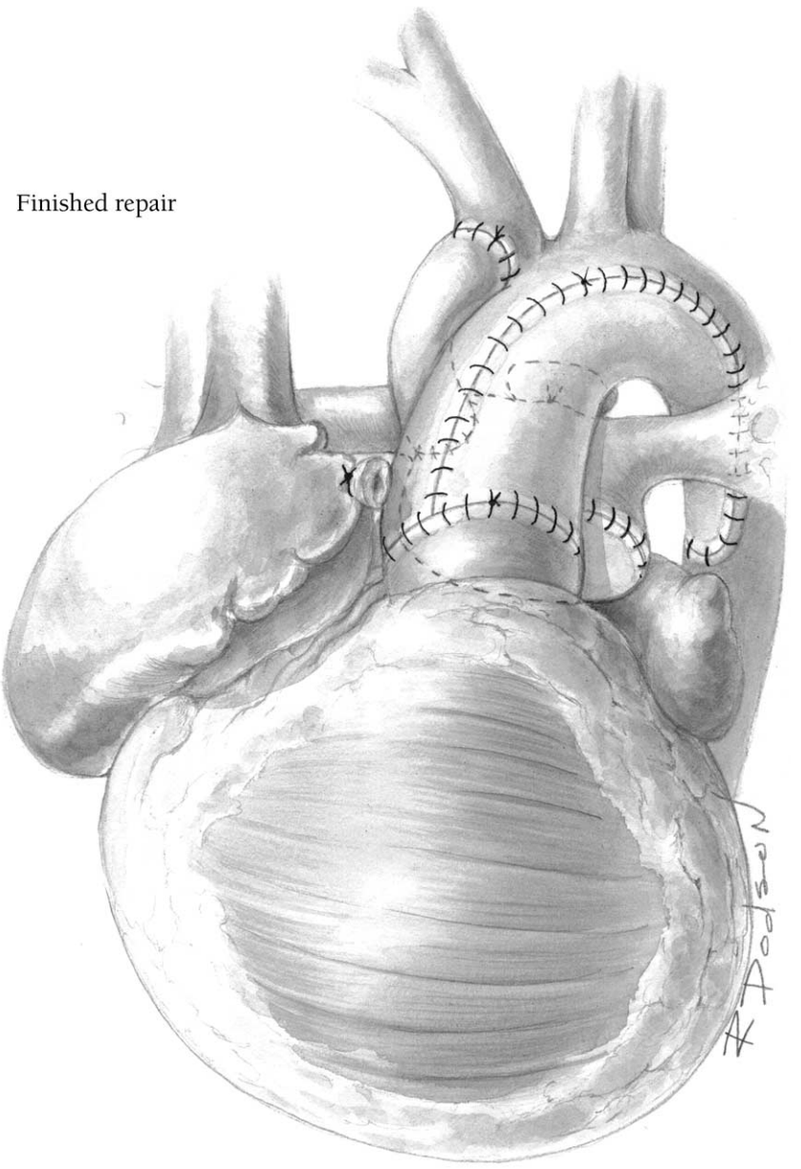

Figure 12 The completed reconstruction.

As rewarming progresses after completion of the reconstruction, low-dose inotropic support with dopamine 3 μg/kg/min is begun, and generally, a phosphodiesterase inhibitor (milrinone) is administered in the bypass circuit. The patient is weaned from bypass and modified ultrafiltration is performed. Milrinone is continued to reduce systemic vascular resistance and to promote systemic perfusion. In the majority of cases no additional inotropic support is required, although these agents are used if ventricular function is significantly diminished. The chest is closed in the majority of cases (approximately 75%) if bleeding. Routine paralysis is not continued beyond the first postoperative night, to avoid delay in extubation and in diuresis. Weaning of inspired oxygen to nontoxic levels is generally accomplished rapidly. Ventilation is maintained at relatively high tidal volumes to prevent atelectasis and alveolar hypoxemia. Patients are generally allowed to awaken on the first postoperative morning and are extubated as soon as possible. Milrinone is transitioned to captopril, and aspirin is administered in an effort to prevent shunt thrombosis. Recently a low-dose heparin infusion has been utilized starting the night of surgery and continuing until enteral administration of aspirin is tolerated.

Results

The outcome of patients undergoing Stage 1 palliation for HLHS at the Children's Hospital of Philadelphia has been reported previously. In a cohort of 102 patients with HLHS, operative survival was 78%, and 1-year survival was 66%. For survivors of the stage 1 operation, 1-year survival was 86%.[12] Risk factors for mortality included a low birth weight (<2.5 kg), the presence of associated cardiac anomalies, the need for mechanical circulatory support, and the presence of a genetic syndrome or extracardiac anomaly. In the absence of a low birth weight and cardiac or extracardiac anomaly, the operative survival was 88%. This is consistent with reports from other institutions.[13] Early mortality is now related primarily to noncardiac abnormalities such as necrotizing en-

terocolitis, neurologic dysfunction, or infection. Thus hospital survival for patients with HLHS now approaches that of many other forms of critical congenital heart disease. Whether this survival will be further increased by any of the technical modifications being proposed now, or by fetal diagnosis and intervention, remains to be elucidated.

As survival following the Stage 1 operation has improved, there has been increasing interest in the neurodevelopmental outcome in patients with HLHS. There is ample evidence that these patients are at risk for neurodevelopmental delay; this can potentially be traced to preoperative decreased cerebral blood flow and organ dysfunction,[14] operative factors such as circulatory arrest or low flow bypass, or postoperative events of hemodynamic instability and chronic cyanosis.[6] Motor skills and attention seem to be at greater risk than verbal skills. The majority of the children remain in the low normal IQ range. The presence of genetic syndrome or perioperative neurologic dysfunction as evidenced by a seizure disorder most strongly portends delayed neurodevelopmental outcome. It is hoped that as techniques for perioperative management and for surgical correction are further refined, improvement in neurological outcome will continue.

References

1. Noonan JA, Nadas AS: The hypoplastic left heart syndrome: an analysis of 101 cases. Pediatric Clin North Am 5:1029, 1958
2. Jacobs JP: Nomenclature and Classification for Congenital Cardiac Surgery, in Mavroudis C, Backer C (eds): Pediatric Cardiac Surgery (ed 3). Philadelphia, PA, Mosby, 2003
3. Norwood WI, Lang P, Hansen DD: Physiologic repair of aortic atresia–hypoplastic left heart syndrome. N Engl J Med 308:23, 1983
4. Bailey LL, Nehlsen-Cannerella SL, Doroshow RW, et al: Cardiac allotransplantation in newborns as therapy for HLHS. N Engl J Med 315: 949, 1986
5. Spray TL: Hypoplastic left heart syndrome, in Gardner TJ, Spray TL (eds): Operative Cardiac Surgery (ed 5). London, Arnold, 2004
6. Mahle WT, Wernovsky G: Neurodevelopmental outcomes in hypoplastic left heart syndrome. Seminars in Thoracic and Cardiovascular Surgery, Pediatric Cardiac Surgery Annual, 2003
7. Wypij D, Newburger J, Rappaport LA, et al: The effect of duration of deep hypothermic circulatory arrest in infant heart surgery on late neurodevelopment: The Boston Circulatory Arrest Trial. J Thorac Cardiovasc Surg 126:1397, 2003
8. Ungerleider RM, Gaynor JW: The Boston Circulatory Arrest Study: an analysis. J Thorac Cadiovasc Surg 127:1256, 2004
9. Bellinger DC, Wypij D, et al: Neurodevelopmental status at eight years in children with dextro-transposition of the great arteries: The Boston Circulatory Arrest Trial. J Thorac Cardiovasc Surg 126:1385, 2003
10. Bove EL, Ohye RG, Devaney EJ: Hypoplastic Left Heart Syndrome: conventional surgical management. Seminars in Thoracic and Cardiovascular Surgery, Pediatric Cardiac Surgery Annual, 2003
11. Sano S, Ishino K, Kawada M, et al: Right ventricle-pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 126:504, 2003
12. Gaynor JW, Mahle WT, Cohen MI, et al: Risk factors for mortality after the Norwood procedure. Eur J Cardiothorac Surg 22:82, 2002
13. Bove EL, Lloyd TR: Staged reconstruction for hypoplastic left heart syndrome: contemporary results. Ann Surg 224:387, 1996
14. Mahle WT, Clancy RR, McGaurn SP, et al: Neurodevelopmental outcome and lifestyle assessment in school-aged and adolescent children with hypoplastic left heart syndrome. Pediatrics 105:1082, 2000