Operative Techniques in Thoracic and Cardiovascular Surgery

Right Ventricle-Pulmonary Artery Shunt in First-Stage Palliation of Hypoplastic Left Heart Syndrome

Shunji Sano, MD, Kozo Ishino, MD, Masaaki Kawada, MD, and Osami Honjo, MD

Hypoplastic left heart syndrome (HLHS) is a spectrum of syndromes displaying underdevelopment of the left-sided heart structure, characterized by aortic valve atresia or severe stenosis with a small ascending aorta and hypoplasia of the left ventricle.[1] Without surgical treatment, practically all infants with this complex cardiac anomaly die within the first month of life.[2,3] Since the introduction of successful surgical palliation in 1981 by Norwood and coworkers,[4] survival of children with HLHS has improved by incremental advances in staged reconstructive surgery for the last two decades.[5-8]

With increased experiences with the Norwood stage I palliation, however, hemodynamic disadvantages associated with a systemic-pulmonary shunt (SP shunt) such as a coronary hypoperfusion due to diastolic runoff and sustained volume overload on the systemic right ventricle[9,10] have been suggested to be related to early and late mortality.[11,12] A review of 122 postmortem cases after the Norwood procedure at the Children's Hospital Boston[9] indicated that the most prevalent causes of death were the impairment of coronary perfusion, excessive pulmonary blood flow, and obstruction to pulmonary blood flow. This maldistribution of blood flow was implicated in more than 60% of postoperative mortality.

Although several institutions with a large surgical volume have achieved early survival rates of 68 to 94% following the Norwood procedure (stage I palliation),[6,7,8] this procedure has still remained a challenging step with unacceptably high mortality for many less experienced centers.[13,14]

History of Right Ventricle-Pulmonary Artery (RV-PA) Shunt in First-Stage Palliation of HLHS

The RV-PA shunt to reestablish pulmonary blood supply in stage I palliation for HLHS was first introduced by Norwood and coworkers in 1981.[4] The shunt materials they used were relatively large for neonates: 8-mm nonvalved polytetrafluoroethylene (PTFE) tubes in two patients and 12-mm valved conduits in another two. These patients all died within 11 hours after surgery either from excessive pulmonary blood flow or from right ventricular failure. In Japan, Kishimoto and coworkers[15] revived the RV-PA shunt procedure using a xenopericardial valved conduit in seven patients and reported stable postoperative hemodynamics with a high diastolic blood pressure. However, their long-term results were unsatisfactory and no other surgeons used the procedure.

The above findings led us to attempt a RV-PA shunt, with the difference that we used a small nonvalved PTFE conduit in first-stage palliation. We constructed a nonvalved PTFE shunt between the right ventricle and the pulmonary artery as an alternative pulmonary blood source. This first attempt was performed on an 8-day-old girl with aortic atresia, mitral atresia, and hypoplasia of ascending aorta using a 4-mm PTFE graft, on February 9, 1998. This patient survived the surgery; however, she developed sudden severe cyanosis 4 months after the surgery and died. A second attempt was performed on February 15, 1998, on a 17-day-old boy with aortic atresia and a mitral atresia. He underwent second-stage bidirectional Glenn anastomosis at 4 months of age and successfully completed a third-stage Fontan operation at 2.8 years of age. Subsequently, we have employed the RV-PA shunt for all the patients with HLHS, although using a small nonvalved PTFE conduit.[16]

In this article we describe our developing surgical techniques over 7 years' institutional experience with a modified Norwood procedure using a RV-PA shunt as stage I palliation for HLHS.

Patients and Methods

Patient Population

Between February 1998 and August 2004, 40 consecutive infants (25 boys and 15 girls) with HLHS underwent a modified Norwood procedure using a RV-PA shunt at Okayama University Hospital. Age at operation ranged from 3 to 57 days (median, 10 days). Weight at operation ranged from 1.6 to 3.9 kg (median, 2.8 kg) and five patients weighed less than 2.0 kg.

Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine and Dentistry, Okayama, Japan.

Address reprint requests to Shunji Sano, MD, Department of Cardiovascular Surgery, Okayama University Graduate School of Medicine and Dentistry, 2-5-1 Shikata-cho, Okayama City 700-8558, Japan; e-mail: s_sano@cc.okayama-u.ac.jp.

doi:10.1053/j.optechstcvs.2005.06.002

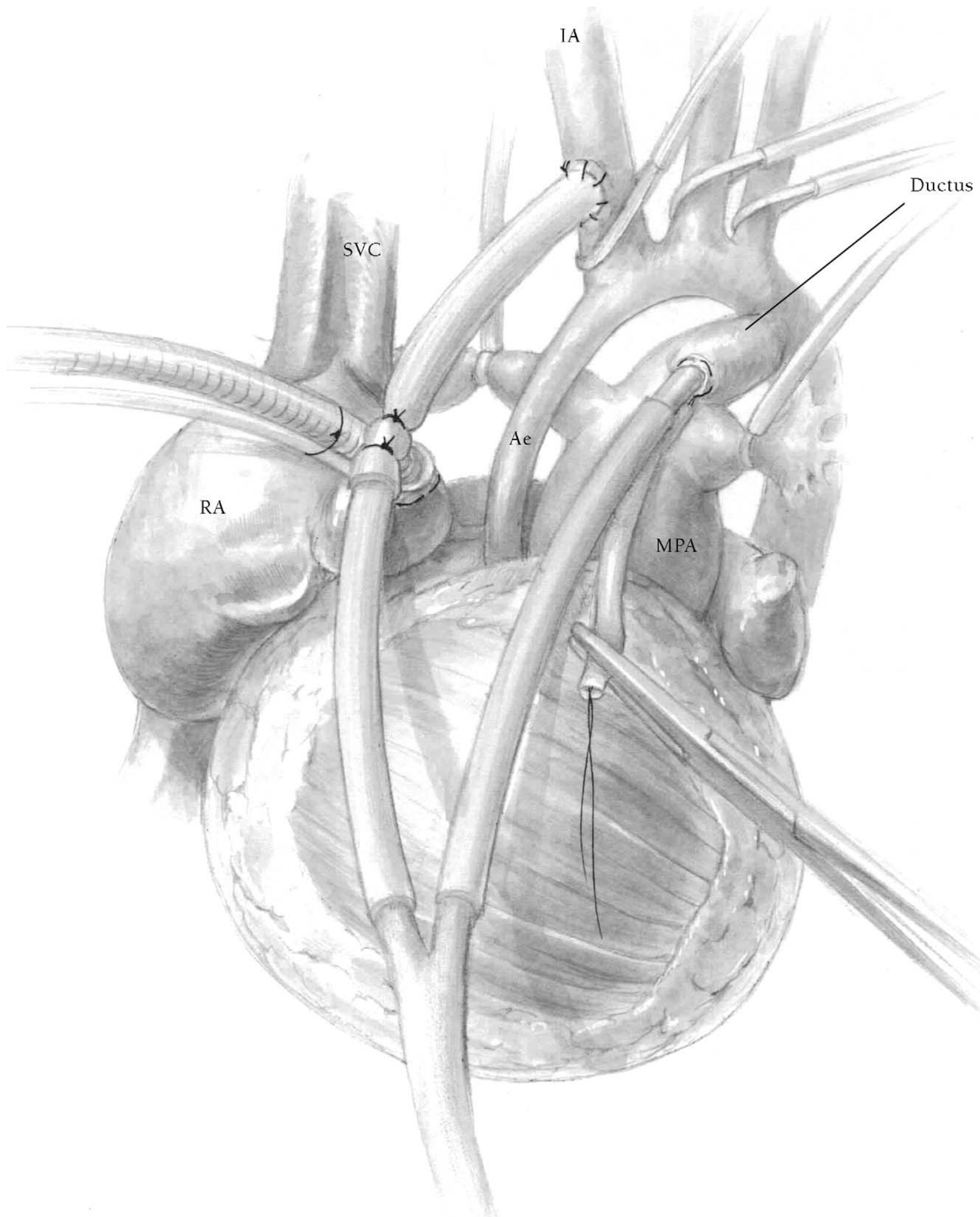

Figure 1 Arterial blood pressure monitoring lines were placed in the right radial artery and femoral artery in each patient preoperatively. Through a midline sternotomy, the thymus gland was excised. The aortic arch, its branches, and the ductus arteriosus were dissected out. To avoid the use of total circulatory arrest as much as possible, cardiopulmonary bypass was established by dual arterial perfusion and single atrial or bicaval drainage cannulations. Dual arterial cannulas were inserted into the ductus arteriosus and into a 3.0-mm polytetrafluoroethylene (PTFE) tube (Gore-Tex, WL Gore & Associates, Inc., Flagstaff, AZ) that was anastomosed to the innominate artery. Following insertion of a venous cannula into the right atrium, cardiopulmonary bypass was commenced at a flow rate of 150 to 180 mL/min/kg.

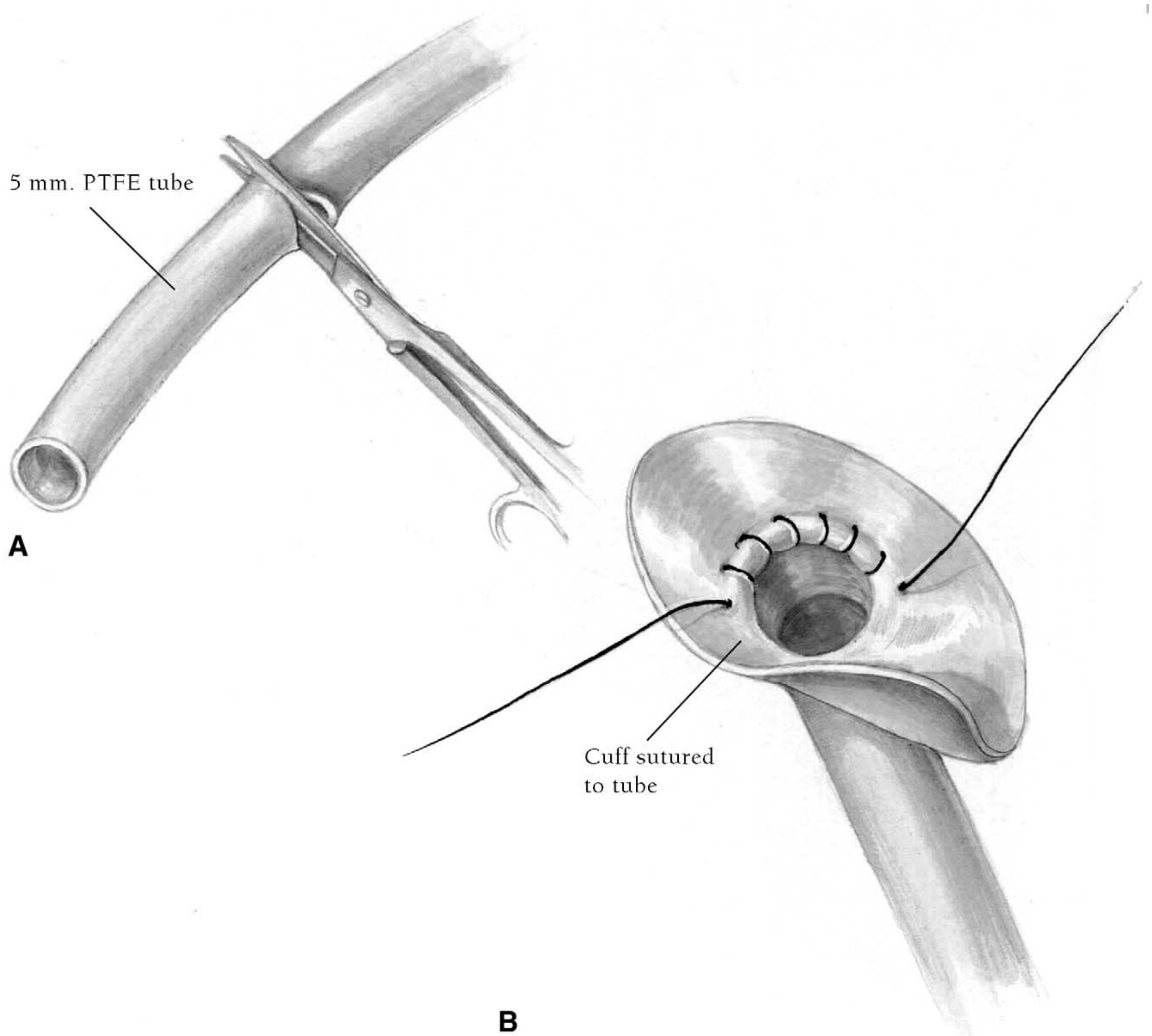

Figure 2 Before establishment of cardiopulmonary bypass, the shunt material was prepared on another table as follows. In the case of 5-mm shunt, a 5-mm PTFE tube was cut in an appropriate length as a shunt tube. A cuff for an anastomosis to the distal end of the main pulmonary artery was tailored by vertically opening a piece of the same PTFE tube. The center of the cuff was punched out in 5-mm diameter, and the tube was anastomosed to this opening using a 8-O PTFE or Prolene suture.

Operative Procedure

Arterial blood pressure monitoring lines were placed in the right radial artery and femoral artery in each patient preoperatively. Through a midline sternotomy, the thymus gland was excised. The aortic arch, its branches, and the ductus arteriosus were dissected out. To avoid the use of total circulatory arrest as much as possible, cardiopulmonary bypass was established by dual arterial perfusion and single-atrial or bicaval drainage cannulations. Dual arterial cannulas were inserted into the ductus arteriosus and into a 3.0-mm PTFE tube (Gore-Tex, WL Gore & Associates, Inc., Flagstaff, AZ) that was anastomosed to the innominate artery. Following insertion of a venous cannula into the right atrium, cardiopulmonary bypass was commenced at a flow rate of 150 to 180 ml/min/kg.

Before establishment of cardiopulmonary bypass, the shunt material was prepared on another table as follows. In the case of a 5-mm shunt, a 5-mm PTFE tube was cut in an appropriate length as a shunt tube. A cuff for an anastomosis to the distal end of the main pulmonary artery was tailored by vertically opening a piece of the same PTFE tube. The center of the cuff was punched out in 5-mm diameter, and the tube was anastomosed to this opening using an 8-O PTFE or Prolene suture.

During the cooling phase, the isthmus was ligated and divided to obtain a better operative field. Following division of the duct proximal to the cannulation site, the descending thoracic aorta was extensively mobilized by blunt dissection as far distally as possible. The main pulmonary artery was transected just proximal to the bifurcation, and the PTFE cuff was anastomosed to the distal stump of the main pulmonary artery using either 8-O PTFE or 8-O Prolene suture.

Although we repaired the distal end of the main pulmonary artery with an autologous pericardial patch and anastomosed the shunt tube onto this patch in the first six patients,

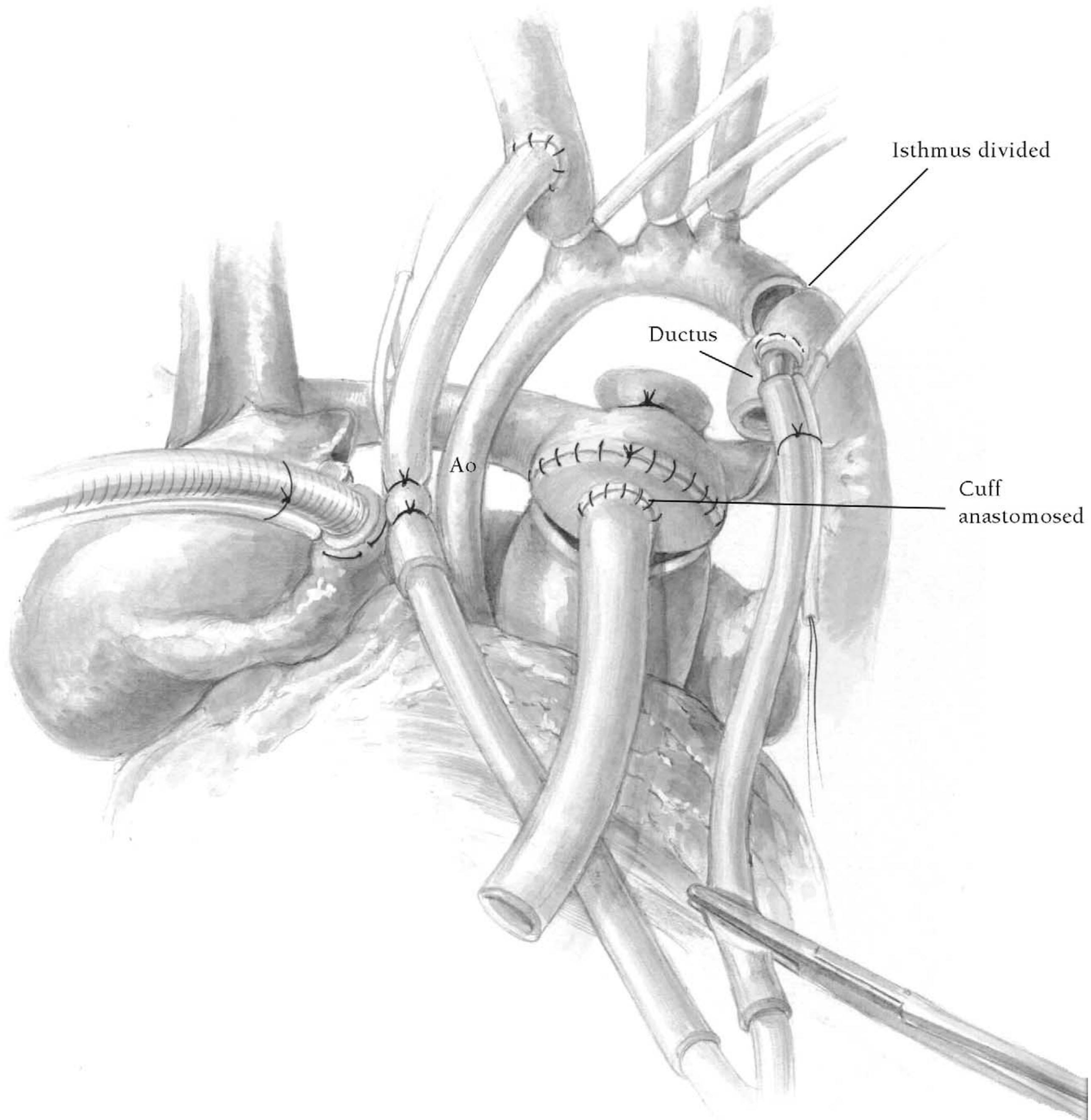

Figure 3 During the cooling phase, the isthmus was ligated and divided to obtain a better operative field. Following division of the duct proximal to the cannulation site, the descending thoracic aorta was extensively mobilized by blunt dissection as far distally as possible. The main pulmonary artery was transected just proximal to the bifurcation, and the PTFE cuff was anastomosed to the distal stump of the main pulmonary artery using either 8-O PTFE or 8-O Prolene suture.

Although we repaired the distal end of the main pulmonary artery with an autologous pericardial patch and anastomosed the shunt tube onto this patch in the first six patients, we abandoned this procedure because of intractable bleeding and the high incidence of late obstruction at the anastomotic site.

we abandoned this procedure because of intractable bleeding and the high incidence of late obstruction at the anastomotic site.

At a nasopharyngeal temperature of less than 22°C, the descending aorta was clamped. Following removal of the perfusion cannula from the duct, all duct tissue was excised from the descending aorta. The left carotid artery and left subclavian artery were snared. Isolated cerebral and myocardial perfusion was established by placing a clamp just distal to the innominate artery.[14,15] With the heart beating, the aortic arch was opened inferiorly and the back wall of the descending aorta was anastomosed to the posterior wall of the aortic arch. At this stage, cold crystalloid cardioplegic solution (30 ml/kg) was administered over 3 minutes either from the aortic root or from a side port of the arterial cannula during temporary total circulatory arrest.

The innominate artery was snared proximal to the perfusion site, and the clamp on the arch was removed. Cardio-

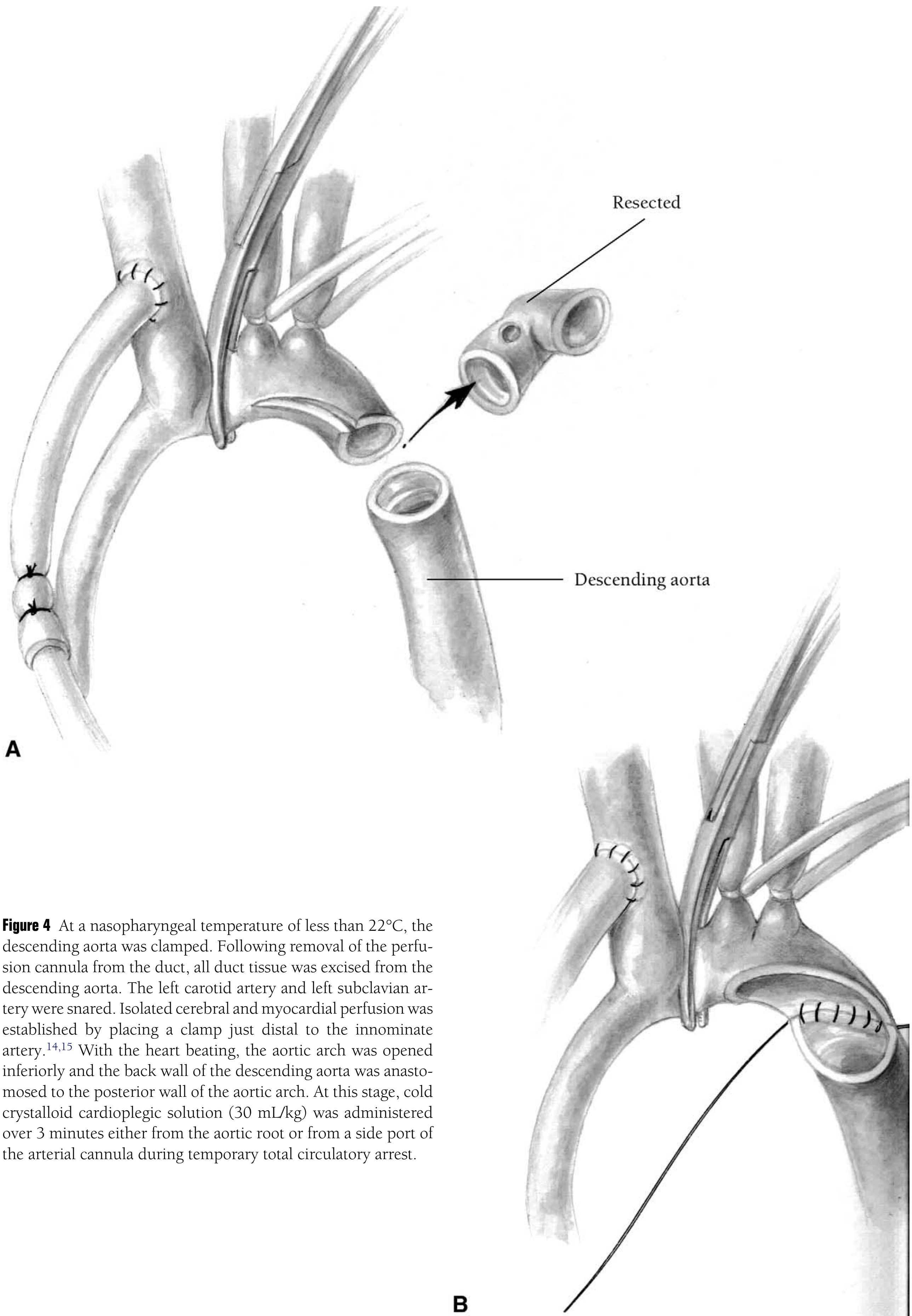

Figure 4 At a nasopharyngeal temperature of less than 22°C, the descending aorta was clamped. Following removal of the perfusion cannula from the duct, all duct tissue was excised from the descending aorta. The left carotid artery and left subclavian artery were snared. Isolated cerebral and myocardial perfusion was established by placing a clamp just distal to the innominate artery.[14,15] With the heart beating, the aortic arch was opened inferiorly and the back wall of the descending aorta was anastomosed to the posterior wall of the aortic arch. At this stage, cold crystalloid cardioplegic solution (30 mL/kg) was administered over 3 minutes either from the aortic root or from a side port of the arterial cannula during temporary total circulatory arrest.

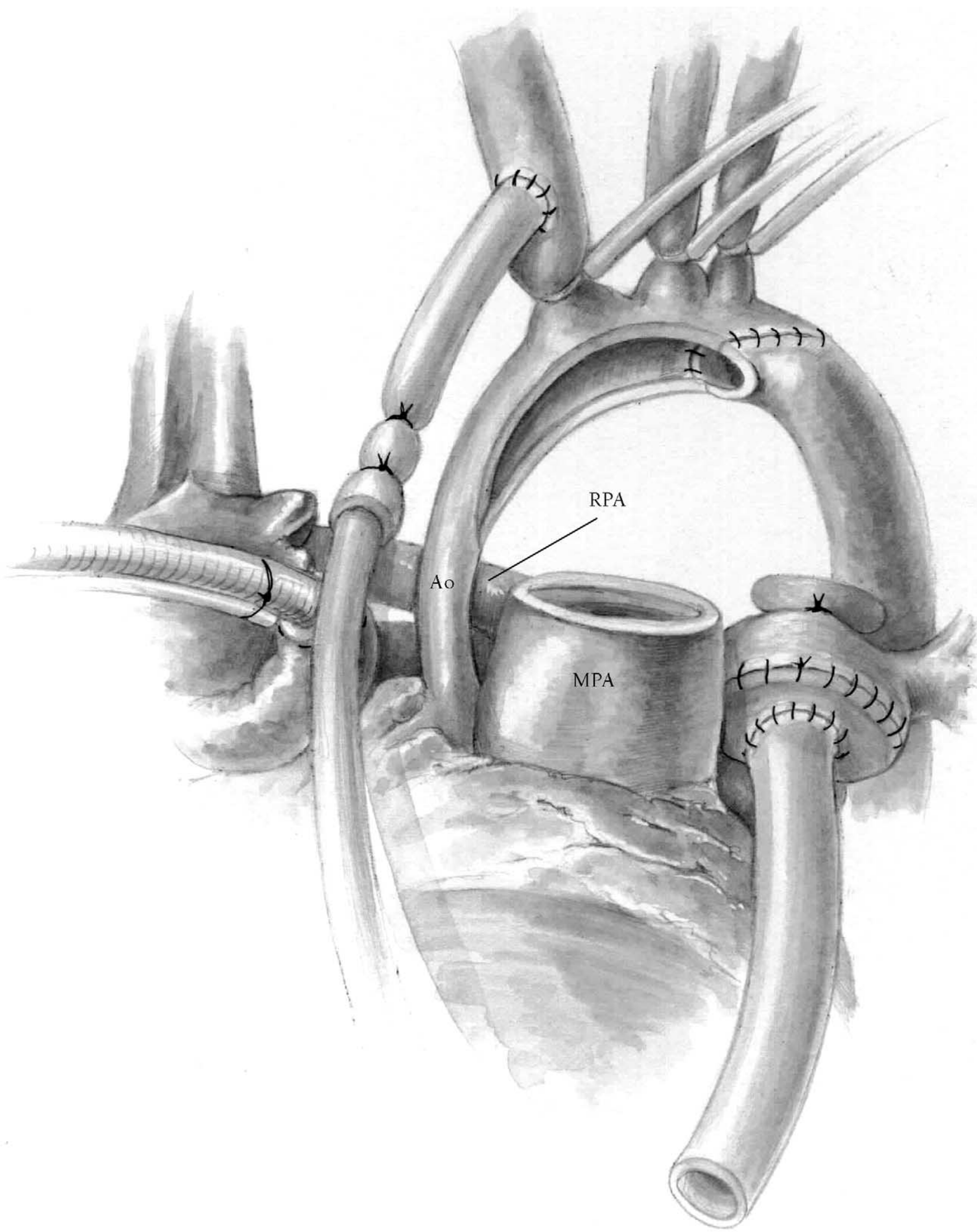

Figure 5 The innominate artery was snared proximal to the perfusion site, and the clamp on the arch was removed. Cardiopulmonary bypass was resumed for isolated cerebral perfusion through the innominate artery. The aortic arch reconstruction was performed by means of Brawn's modification.[3]

pulmonary bypass was resumed for isolated cerebral perfusion through the innominate artery. The aortic arch reconstruction was carried out by means of Brawn's modification.[3] The opening of the aortic arch was extended down into the ascending aorta to the level of the transected end of the main pulmonary artery. The proximal main pulmonary artery was directly anastomosed to the transverse arch and the opened-out ascending aorta.

After completion of aortic arch reconstruction, the circulation was again arrested. The venous cannula was removed from the right atrium. The atrial septum was excised, working through the purse-string suture on the atrial appendage.

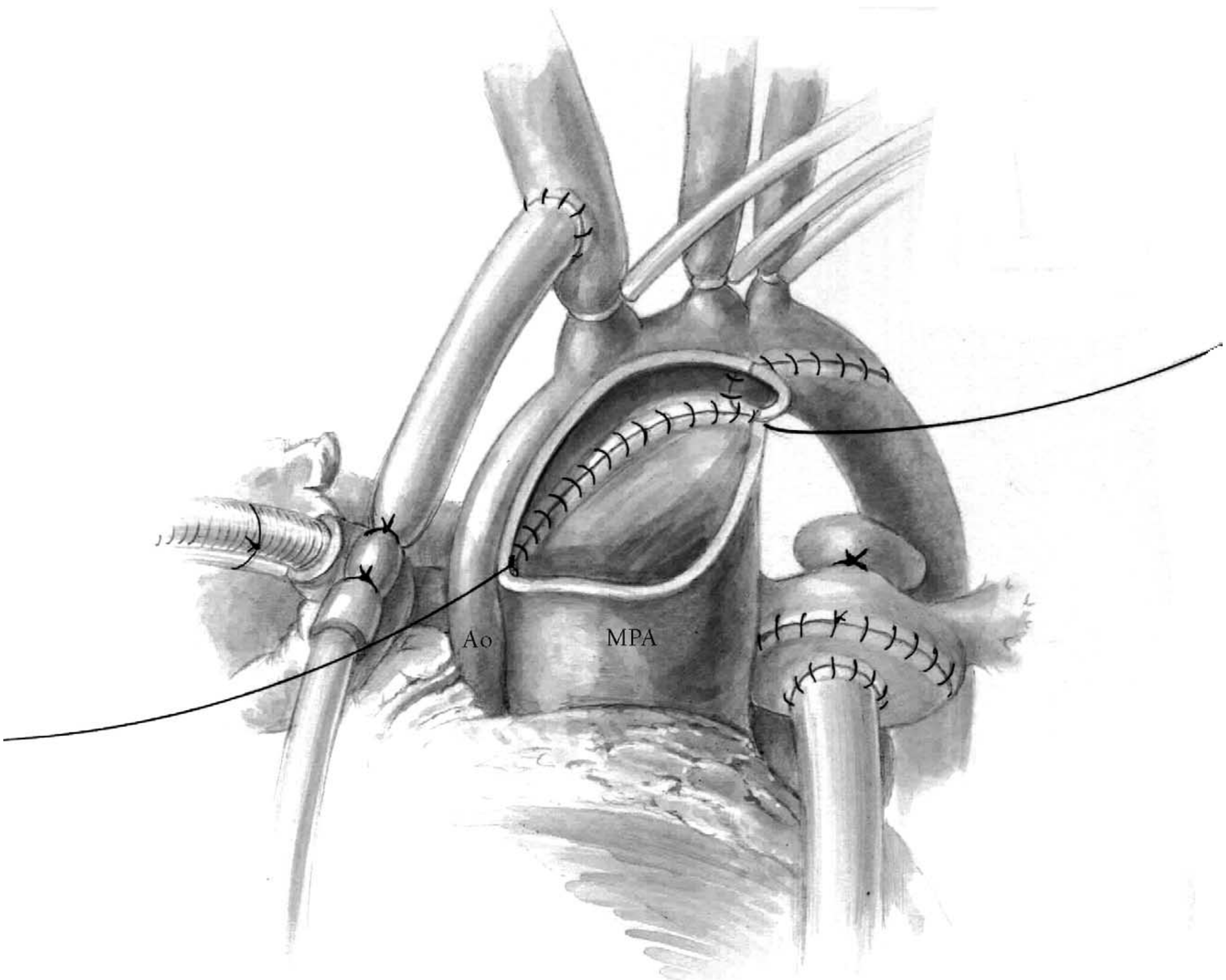

Figure 6 The opening of the aortic arch was extended down into the ascending aorta to the level of the transected end of the main pulmonary artery. The proximal main pulmonary artery was directly anastomosed to the transverse arch and the opened-out ascending aorta.

An appropriate site of ventriculotomy for the RV-PA shunt, which was 1.5 to 2 cm below the pulmonary valve, was made on the right ventricular outflow tract. To prevent late obstruction at the shunt anastomosis, it was important to punch out a piece of ventricular muscle underlying the ventriculotomy. The size of the ventricular hole is approximately 1 mm larger than the PTFE graft.

Cardiopulmonary bypass was reinstituted, and all snares were removed. The RV-PA shunt was placed to the left of the neoaorta in all patients and was anastomosed to the right ventriculotomy during rewarming using 6-O prolene suture. The size of the RV-PA shunt used was 4 mm in five patients weighing less than 2.0 kg, 6 mm in one patient with a weight of 3.7 kg, and 5 mm in the other patients. Modified ultrafiltration was routinely used following weaning from cardiopulmonary bypass.

Postoperative Management

The sternum was routinely left open and patients underwent delayed sternal closure on postoperative day 1 to 11 (median, 3 days). Postoperative intensive care of patients with the RV-PA shunt was basically the same as that for neonates undergoing other types of surgery. Thus, delicate manipulations to control pulmonary and systemic vascular resistance were not necessary. Ventilator settings were adjusted to keep arterial oxygen saturations higher than 75% and carbon dioxide levels lower than 45 mm Hg. It should be kept in mind that hypercarbia, which is often used to increase pulmonary vascular resistance in patients with the systemic-pulmonary shunt, can easily cause hypoxemia and subsequent hemodynamic deterioration in those with the RV-PA shunt. The inotropic drugs most used were dopamine or dobutamine 5 μg/kg per minute and epinephrine 0.05 to 0.1 μg/kg per minute, and, if necessary, calcium chloride 0.25 mmol/h was also used.

Results

Stage I Norwood Procedure

All patients were weaned from CPB. Hemodynamic instability did not occur in any patients, and there were no special ventilatory manipulations.

There were 37 hospital survivors (92%), including five

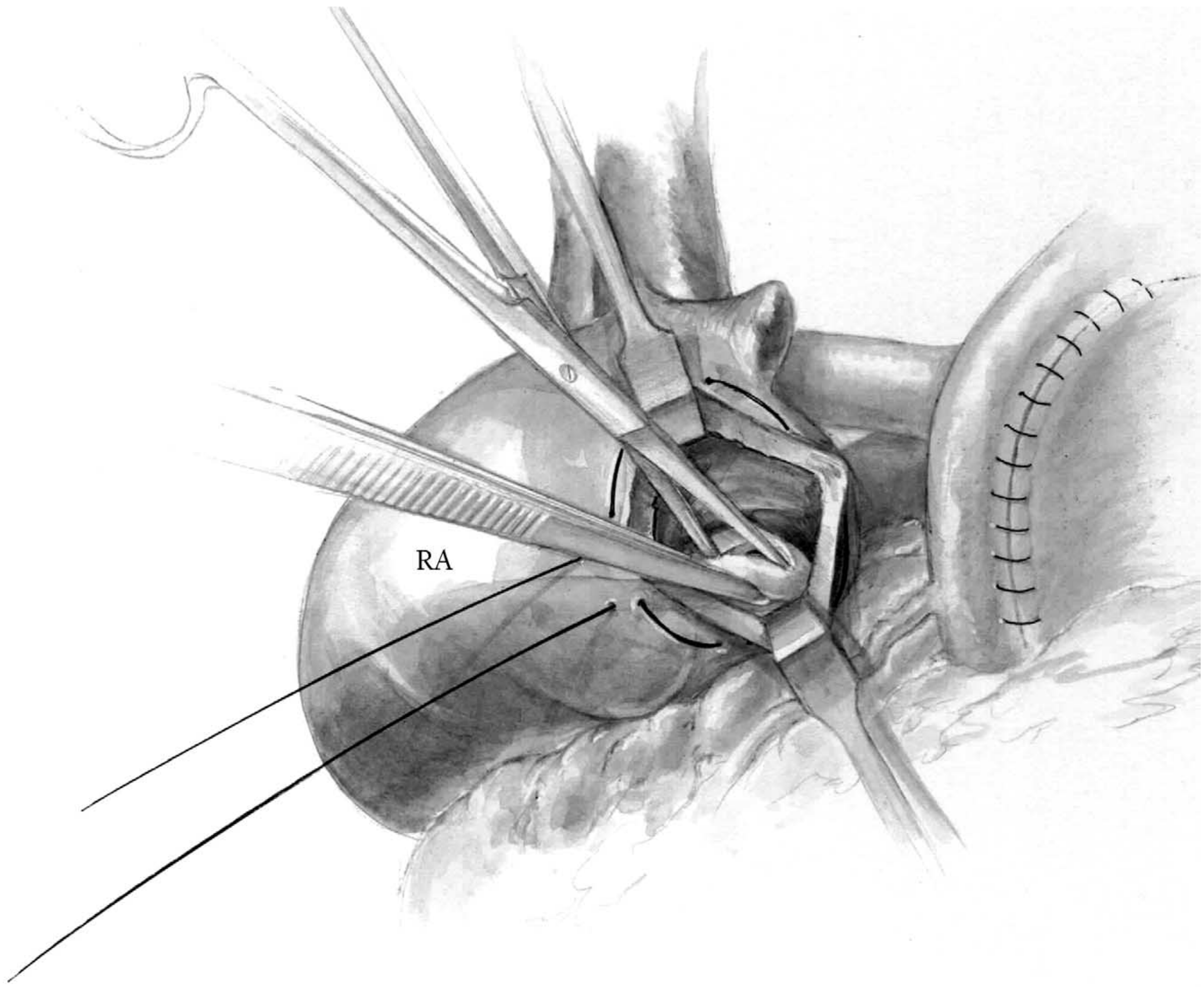

Figure 7 After completion of aortic arch reconstruction, the circulation was again arrested. The venous cannula was removed from the right atrium. The atrial septum was excised, working through the purse-string suture on the atrial appendage.

patients weighing less than 2.0 kg. One patient died from sudden cardiac arrest on the next postoperative day, and one other patient died from septicemia 2 weeks after the operation. There were two late deaths among the first four patients. Both died from severe hypoxemia due to obstruction of the RV-PA shunt (3 and 4 months after surgery).

Twenty-five patients underwent BDG (mean age, 5.7 months; median, 5.6 months; range, 1.1 to 11.3 months). There were two hospital deaths: one from viral pneumonia and the other from progressive hypoxia, and no late deaths.

The Fontan operation has been performed at a median age of 2.4 years (mean, 2.5 years; range, 1.8 to 3.2 years). There were no hospital or late mortalities.

Comment

Despite successful reconstructive surgery, most deaths occur in the first 24 to 48 hours after surgery due to hemodynamic instability secondary to the unpredictable rapid fall in pulmonary vascular resistance. Over the last decade, therefore, many efforts to achieve a balanced circulation have focused on limiting pulmonary blood flow and increasing systemic oxygen delivery. These measures have included reduction in the size of the shunt,[5,8] use of systemic vasodilators,[7,17] and induction of hypoxemia and hypercarbia by ventilator manipulations[6,7,8,18] and/or administration of hypoxic admixtures.[6,17] Such delicate postoperative management, however, requires much manpower on a 24-hour basis.

The fundamental advantages of RV-PA shunt were that elimination of diastolic runoff into the pulmonary circulation resulted in high diastolic pressure, which was constantly over 40 mm Hg throughout the entire postoperative period, and coronary perfusion was independent of the pulmonary-to-systemic flow ratio. Independent coronary perfusion resulted in a very stable postoperative course.[19]

The necessity for a right ventriculotomy, which may affect contractile function of the systemic ventricle, is a potential disadvantage of the RV-PA shunt. Norwood and colleagues[4] attempted to use the RV-PA shunt in stage I palliation with relatively large materials, 8-mm nonvalved PTFE tube, or 12-mm valved conduit; however, all the patients died due to excessive pulmonary blood flow or right ventricular failure. Similar findings were observed in Kishimoto's report.[15]Thus, a large ventriculotomy in neonates will surely impair right ventricular function. Another concern related to a ventricu-

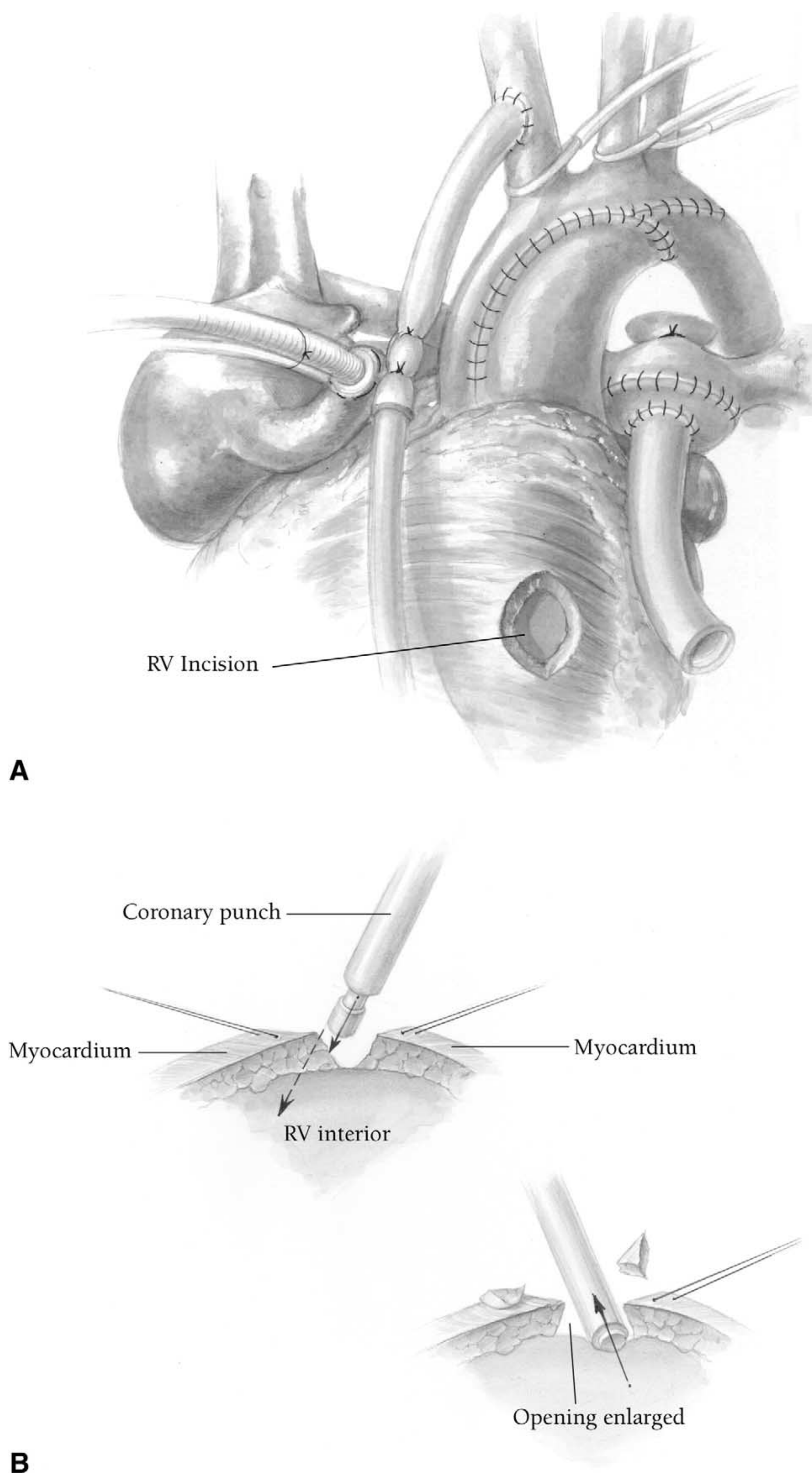

Figure 8 An appropriate site of ventriculotomy for the RV-PA shunt, which was 1.5 to 2 cm below the pulmonary valve, was made on the right ventricular outflow tract. In order to prevent late obstruction at the shunt anastomosis, it was important to punch out a piece of ventricular muscle underlying the ventriculotomy. The size of the ventricular hole is approximately 1 mm bigger than PTFE graft.

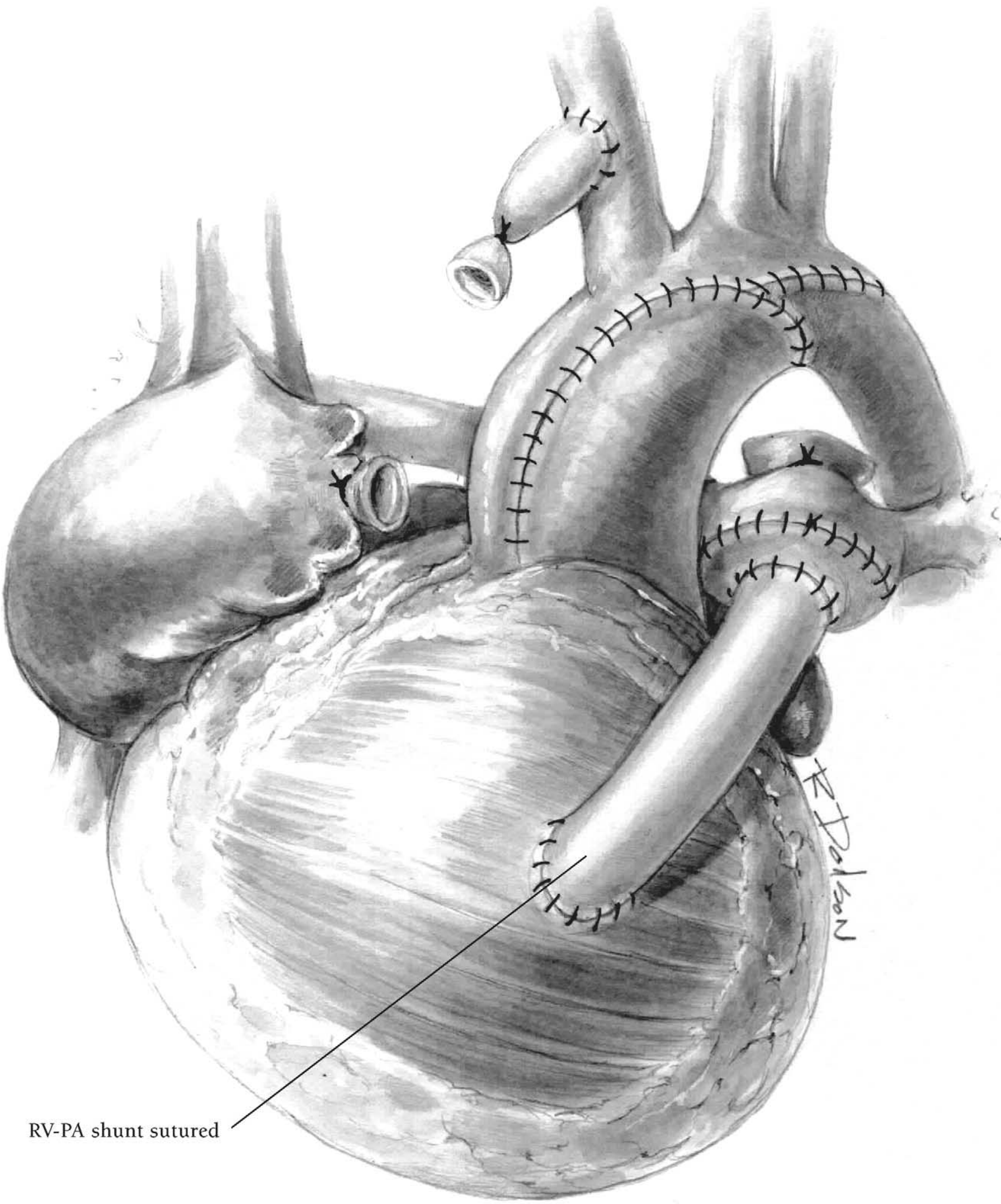

Figure 9 Cardiopulmonary bypass was reinstituted, and all snares were removed. The RV-PA shunt was placed to the left of the neoaorta in all patients and was anastomosed to the right ventriculotomy during rewarming using 6-0 prolene suture. The size of the RV-PA shunt used was 4 mm in five patients weighing less than 2.0 kg, 6 mm in one patient with a weight of 3.7 kg, and 5 mm in the other patients. Modified ultrafiltration was routinely used following weaning from cardiopulmonary bypass.

lotomy is complication of ventricular arrhythmia. Follow-up studies in patients with tetralogy of Fallot identified ventriculotomy as an incremental risk factor for late arrhythmias.[20,21] Our results indicate that a small right ventriculotomy made in the upper outflow tract for an anastomosis with a 4- to 6-mm conduit exerts little influence on ventricular function and does not become arrhythmogenic at least until Fontan completion[22]; however, long-term follow-up is necessary.

The lessons learned from our 7 years of experience with the modified Norwood procedure using the RV-PA shunt are twofold. First, the nonvalved PTFE RV-PA shunt becomes obstructive over time, particularly 3 months or more after surgery. Since two patients in the early series died from progressive shunt obstruction after hospital discharge, this prompted us to check the patients' saturations every week at an outpatient clinic. When patients present with progressive desaturation, cardiac catheterization should be performed as soon as possible. Our current strategy first involves attempting to perform balloon angioplasty for significant stenoses in the RV-PA shunt or the branch pulmonary arteries. If angioplasty fails or the patient's saturation continues to drop, we then perform the BDG anastomosis. It should also be noted that pulmonary artery growth in patients who received the RV-PA shunt is inadequate, as demonstrated in the patients undergoing the Fontan operation. However, our results indicate that lower pulmonary vascular resistance makes subsequent Fontan procedures feasible even in patients with small pulmonary arteries.

In conclusion, the RV-PA shunt in stage I reconstruction provides stable hemodynamics without requiring extensive postoperative medical intervention. We therefore propose that improvement in the survival of infants with HLHS can be accomplished by many less experienced surgeons by applying the modified Norwood procedure with the RV-PA shunt.

References

1. Noonan JA, Nadas AS: The hypoplastic left heart syndrome: an analysis of 101 cases. Pediatr Clin North Am 5:1026-1056, 1958
2. Sinha SN, Rusnak SL, Sommers HM, et al: Hypoplastic left ventricle syndrome: analysis of thirty autopsy cases in infants with surgical consideration. Am J Cardiol 21:166-173, 1968
3. Hawkins JA, Doty DB: Aortic atresia: morphologic characteristics affecting survival and operative palliation. J Thorac Cariovasc Surg 88: 620-626, 1984
4. Norwood WI, Lang P, Castaneda AR, et al: Experience with operation for hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 82:511-519, 1981
5. Bove EL, Lloyd TR: Staged reconstruction for hypoplastic left heart syndrome. Ann Surg 224:387-395, 1996
6. Mahle WT, Spray TL, Wernovsky G, et al: Survival after reconstructive surgery for hypoplastic left heart syndrome: a 15-year experience from a single institution. Circulation 102:III-136-141, 2000 (suppl III)
7. Tweddell JS, Hoffman GM, Mussatto KA, et al: Improved survival of patients undergoing palliation of hypoplastic left heart syndrome: lessons learned from 115 consecutive patients. Circulation 106:I-81-89, 2002 (suppl I)
8. Ishino K, Stumper O, De Giovanni JJV, et al: The modified Norwood procedure for hypoplastic left heart syndrome: early to intermediate results of 120 patients with particular reference to aortic arch repair. J Thorac Cardiovasc Surg 117:920-930, 1999
9. Bartram U, Grunenfelder J, Van Praagh R: Causes of death after the modified Norwood procedure: a study of 122 postmortem cases. Ann Thorac Surg 64:1795-1802, 1997
10. Mosca RS, Bove EL, Crowley DC, et al: Hemodynamic characteristics of neonates following first stage palliation for hypoplastic left heart syndrome. Circulation 92:II-267-271, 1995 (suppl II)
11. Mahle WT, Spray TL, Gaynor JW, et al: Unexpected death after reconstructive surgery for hypoplastic left heart syndrome. Ann Thorac Surg 71:61-65, 2001
12. Altmann K, Printz BF, Solowiejczyk DE, et al: Two-dimensional echocardiographic assessment of right ventricular function as a predictor of outcome in hypoplastic left heart syndrome. Am J Cardiol 86:964-968, 2000
13. Kern JH, Hayes CJ, Michler RE, et al: Survival and risk factor analysis for the Norwood procedure for hypoplastic left heart syndrome. Am J Cardiol, 80:170-174, 1997
14. Jacobs ML, Blackstone EH, Bailey LL: The Congenital Heart Surgeons Society. Intermediate survival in neonates with aortic atresia: a multi-institutional study. J Thorac Cardiovasc Surg 116:417-431, 1998
15. Kishimoto H, Kawahira Y, Kawata H, et al: The modified Norwood palliation on a beating heart. J Thorac Cardiovasc Surg 118:1130-1132, 1999
16. Sano S, Ishino K, Kawada M, et al: Right ventricle-to-pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg 126:504-510, 2003
17. Jobes DR, Nicolson SC, Steven JM, et al: Carbon dioxide prevents pulmonary overcirculation in hypoplastic left heart syndrome. Ann Thorac Surg 54:150-151, 1991
18. Hoffman GM, Ghanayem NS, Kampine JM, et al: Venous saturation and the anaerobic threshold in neonates after the Norwood procedure for hypoplastic left heart syndrome. Ann Thorac Surg 70:1515-1521, 2000
19. Sano S, Ishino K, Kawada M, et al: Right ventricle-to-pulmonary artery shunt in first stage palliation of hypoplastic left heart syndrome. Semin Thorac Cardiovasc Surg 7:22-31, 2004
20. Gatzoulis MA, Balaji S, Webber SA, et al: Risk factors for arrhythmia and sudden cardiac death later after repair of tetralogy of Fallot: a multicentre study. Lancet, 356:975-981, 2000
21. Dietl CA, Cazzaniga ME, Dubner SJ, et al: Life-threatening arrhythmias and RV dysfunction after surgical repair of tetralogy of Fallot: comparison between transventricular and transatrial approaches. Circulation 90:II-7-12, 1994
22. Sano S, Ishino K, Kawada M, et al: Five year experience with RV-PA shunt in first stage palliation of hypoplastic left heart syndrome—influence to pulmonary artery growth and cardiac function. Ann Thorac Surg (in press)