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Cardiol Young 2004; 14 (Suppl. 3): 90–95

ISSN 1047-9511

Original Article

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Keywords: Hypoplastic left heart syndrome; Norwood; systemic-to-pulmonary shunt

THE HYPOPLASTIC LEFT HEART SYNDROME includes a spectrum of underdevelopment of the left-sided cardiac structures, characterized by aortic valvar 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 Norwood et al.[4] reported successful surgical palliation in 1981, multi-staged reconstructive surgery, involving the Norwood procedure as the first stage, followed by a bi-directional Glenn anastomosis and subsequent completion of the Fontan circulation, has become the established surgical treatment.

Despite successful aortic reconstruction, maldistribution of the cardiac output associated with the systemic-to-pulmonary shunt has been implicated as a major cause of early death after initial palliation.[5,6] Over the last decade, therefore, many efforts have been made to achieve balanced circulations in patients with hypoplasia of the left heart, focusing on the limitation of flow of blood to the lungs, and maximization of systemic delivery of oxygen. These measures have included reduction in the size of the shunt,[6,7] use of systemic vasodilators,[8,9] and induction of hypoxemia and hypercarbia by manipulation of ventilation[6,9–11] and/or hypoxic admixtures.[8,12] In addition, indirect assessment of the cardiac output and systemic delivery of oxygen were generally performed on a 24-hour basis. Since 1998, we have constructed a non-valved polytetrafluoroethylene shunt between the right ventricle and the pulmonary arteries in place of the traditional systemic-to-pulmonary arterial shunt. In this review, we describe the initial 5 years of our institutional experience using this modification of the Norwood procedure.

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Accepted for publication October 2003

Patients and methods

Between February 1998 and February 2003, we treated 27 consecutive infants, 15 boys and 12 girls, with hypoplasia of the left heart by placing a shunt directly from the right ventricle to the pulmonary arteries. Upon arrival at Okayama University Hospital, all patients underwent detailed echocardiography. In 20 patients, we found the classic patterns of hypoplasia of the left heart, including: aortic atresia or stenosis, mitral atresia or stenosis with a poorly developed left ventricle, normally related great arteries, and an intact ventricular septum. The remaining 7 patients had variants of the syndrome with hypoplasia of the left ventricle and aortic arch. In 3, there was double-outlet right ventricle, with aortic and subaortic stenoses. Heterotaxy syndrome was present in 2, both having asplenia with right isomerism and aortic atresia. Aortic and mitral stenosis in presence of a ventricular septal defect were found in 1 patient, and the final patients had aortic atresia with interruption of the aortic arch between the carotid arteries in presence of a ventricular septal defect. Associated anomalies included a persistent left superior caval vein in 5 patients, a vascular sling in 1, totally anomalous pulmonary venous connection in 1, and retrooesophageal course of the right subclavian artery in 1.

The diameter of the ascending aorta ranged from 1.6 to 7.6 millimeters, with a mean of 3.8 millimeters and a median of 3.0 millimeters. The aorta measured 2 millimeters or less in 5 patients. Right ventricular fractional shortening ranged from 24% to 43%, with a mean of 33% and a median of 32%. Tricuspid regurgitation, as documented by color flow Doppler echocardiography, was absent in 5 patients, mild in 11, moderate in 9, and severe in 2.

The ages at the time of operation ranged from 3 to 57 days, with a mean of 13 days, and a median of 10 days. The weights at the time of operation ranged from 1.6 to 3.9 kilograms, with a mean of 2.7 and a median of 2.8 kilograms. Of the patients, 4 weighed less than 2.0 kilograms. Mechanical ventilation was administered before the operation to 20 patients, and all but 1 patient received an infusion of prostaglandin E_1. Peritoneal dialysis was needed in 1 patient due to acute renal failure secondary to ductal shock.

Operative procedure

A detailed procedure for the surgery has been reported elsewhere.[13] We will summarise here key points at each step of the modified Norwood procedure using the shunt placed directly from the right ventricle to the pulmonary arteries.

Cardiopulmonary bypass

To avoid using total circulatory arrest as much as possible, cardiopulmonary bypass was established by dual arterial perfusion and single atrial or bicaval drainage cannulations. One perfusion cannula was inserted into a polytetrafluorethylene tube of 3 millimeters diameter that was anastomosed to the brachiocephalic artery in order to maintain continuous cerebral perfusion in all patients. The other perfusion cannula was inserted into the descending aorta from the arterial duct. During the aortic reconstruction, this cannula was removed, and circulation in the lower body was arrested at between 20 and 22 degrees celsius. In 1 patient, who had suffered from renal failure before the operation, an additional perfusion cannula was directly inserted into the descending aorta in the lower posterior mediastinum to maintain the systemic circulation. Cardioplegic solution was administered from a side port of the perfusion cannula during temporary total circulatory arrest. Modified ultrafiltration was routinely used following weaning from cardiopulmonary bypass.

Aortic reconstruction and atrial septectomy

Because homografts are not available in Japan, we reconstruct the arch without use of a patch.[6] In all patients, ductal tissue was completely excised, and a neoaorta was constructed by direct anastomosis of the descending aorta and the proximal part of the pulmonary trunk to the opened diminutive native aorta. A small piece of pericardial patch was used in some patients. An atrial septectomy was performed during the period of total circulatory arrest for infusion of cardioplegic solution. In 1 patient with right isomerism, we also repaired a common atrioventricular valve during the Norwood procedure.

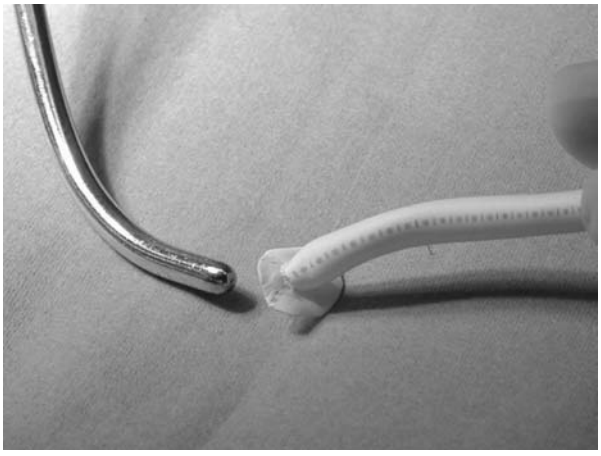

Figure 1.
A 5 millimetre polytetrafluoroethylene tube with a cuff and a 5 millimetre Hegar dilator.

Insertion of the shunt

Before establishing of cardiopulmonary bypass, we prepared the material to be used for the shunt, cutting a polytetrafluorethylene tube of 5 millimeter diameter to the appropriate length. A cuff for an anastomosis to the distal end of the pulmonary trunk was tailored by opening vertically a piece of the same tube. The center of the cuff was punched out to a diameter of 5 millimeters, and the tube was anastomosed to this opening (Fig. 1). During the phase of cooling, the pulmonary trunk was transected just proximal to its bifurcation, and the cuff was anastomosed to the distal stump. For the proximal anastomosis of the shunt, we made a small right ventriculotomy from 1 to 2 centimeters below the pulmonary valve. To prevent late obstruction at the proximal anastomosis, it is of great importance to slice off a piece of ventricular muscle underlying the ventriculotomy. We chose a shunt of 4 millimeter diameter in the first 5 patients, and 5 millimeters diameter in the other patients (Fig. 2).

Postoperative management

The sternum was left open in 24 patients, and closed primarily in 3 patients. Postoperative intensive care

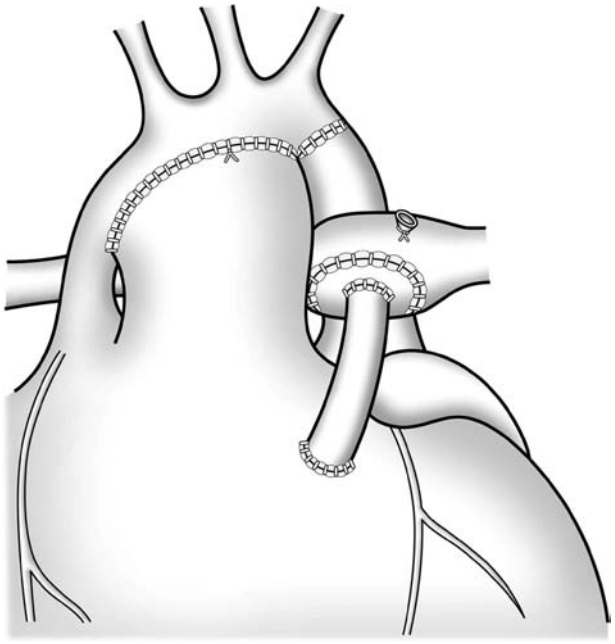

Figure 2.
Completion of the modified Norwood procedure by means of a Damus-type aortic reconstruction and placement of a non-valved conduit from the right ventricle to the pulmonary arteries.

was basically the same as for neonates undergoing other types of surgery. Thus, delicate manipulations to control pulmonary and systemic vascular resistance were not necessary. The ventilator was adjusted to keep arterial saturations of oxygen higher than 75%, and levels of carbon dioxide lower than 45 millimeters of mercury. It should be kept in mind that hypercarbia, which is often used to increase pulmonary vascular resistance in patients with the traditional systemic-to-pulmonary shunt used for initial palliation of hypoplasia of the left heart, easily causes hypoxemia. This could results in subsequent hemodynamic deterioration in those with the shunt placed directly from the right ventricle to the pulmonary arteries. The inotropic drugs we used most were dopamine or dobutamine at a dose of 5 micrograms per kilogram per minute, and epinephrine at between 0.05 and 0.1 micrograms per kilogram per minute. If necessary, calcium chloride at 0.25 millimoles per hour was also used.

Results

The mean time needed for cardiopulmonary bypass during the initial palliation was 157 minutes, with a range from 119 to 254 minutes. The mean myocardial ischemic time was 45 minutes, with a range from 27 to 94 minutes, and the mean renal ischemic time was 48 minutes, with a range from zero to 99 minutes. The mean total circulatory arrest time in 22 patients was 9 minutes, with a range from 3 to 28 minutes. All patients were weaned from bypass. Without the need for any particular ventilatory manipulation, diastolic blood pressure remained above 40 millimeters of mercury, and we did not encounter any hemodynamic instability in any of the patients. There were 25 patients surviving to be discharged from hospital (93%), including 4 infants weighing less than 2 kilograms. In 1 patient, there was a sudden cardiac arrest on the day after surgery. The other patient died from septicemia after 2 weeks. We lost 2 of the first 4 patients subsequent to discharge from hospital. Both died from severe hypoxemia due to obstruction of the shunt, 3 months and 4 months after surgery, respectively. Among the last 18 patients to undergo our procedure, there were no hospital or late deaths before the second stage of reconstruction.

Prior to construction of the bi-directional Glenn anastomosis, 4 patients underwent balloon dilation at either the proximal or distal end of the shunt at the time of cardiac catheterization. We constructed the Glenn anastomosis in 17 patients at a median age of 6.3 months, with a range from 2.6 to 11.3 months. If we found any stenosis related to the shunt, usually seen at the point of anastomosis for the cuff, this was relieved by passage of a 3 or 4 millimeter Hegar dilator through a pulmonary arteriotomy. The shunt itself was left open to serve as an additional supply of blood to the lungs in all patients. The surviving patient with right isomerism underwent simultaneous repair of totally anomalous pulmonary venous connection. In 2 patients with persistence of the left superior caval vein, we constructed anastomoses bilaterally, constructing the right-sided shunt through a median sternotomy and the left-sided one through a lateral thoracotomy after 1 and 3 months, respectively. We lost a further 2 patients after these procedures, 1 from viral pneumonia, and the other from progressive hypoxemia.

We have proceeded to the Fontan operation in 8 patients, constructing a lateral tunnel in 7 and an extracardiac connection in the other. The operations were performed at a median age of 2.6 years, with a range from 1.8 to 3.2 years. The shunt was transected at this stage. In 1 patient, we also replaced the tricuspid valve. The mean pulmonary arterial index,[14] measured on the cineangiogram before the Fontan operation in all patients, was 158, with a range from 140 to 180. The mean right ventricular fractional shortening evaluated by echocardiography after the Fontan operation was 35%, with a range from 30% to 43%. There were no deaths either before or after discharge from hospital.

The current status of all survivors after their first stage reconstruction is shown in Figure 3. The median period of follow-up for the 21 survivors was

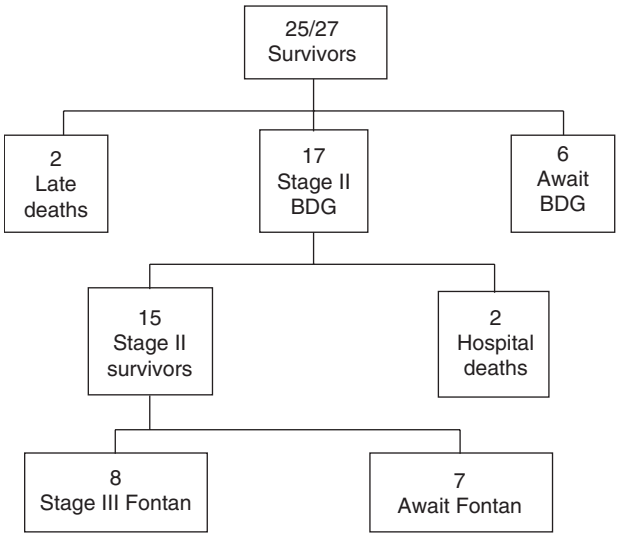

Figure 3.
The current state of our 25 patients who survived the initial palliation and were discharged from hospital.

25 months, with a range from 2 to 63 months. At the time of the last echocardiographic evaluation, tricuspid or common atrioventricular valvar regurgitation was absent or trivial in 11 patients, mild in 7, moderate in 2 and severe in 1. The actuarial survival rate 1 year after the operations of the 27 patients who underwent the modified Norwood procedure using the shunt placed directly from the right ventricle to the pulmonary arteries was 75%, with 95% confidence intervals between 57% and 93%. No deaths have occurred thereafter.

Discussion

The purpose of the initial stage of palliation for hypoplasia of the left heart is to provide unobstructed flow of systemic blood from the right ventricle to the aorta and coronary arteries, to reestablish a supply of blood to the lungs, and to relieve obstruction to pulmonary venous return through the atrial septum. Norwood et al. initially used a valved or non-valved conduit placed from the right ventricle to the pulmonary arteries in the first 4 infants while developing their reconstructive operation for palliation of the hypoplastic left heart syndrome.[4] These patients all died soon after surgery, either from excessive pulmonary blood flow or from right ventricular failure.[15] Kishimoto et al.[16] revived this technique using a valved xenopericardial roll. Since February 1998, we have employed the shunt placed from the right ventricle directly to the pulmonary arteries, albeit employing a small non-valved conduit of polytetraflourethylene. Our experience has clearly shown differences in the postoperative hemodynamics between the direct shunt and the more traditional systemic-to-pulmonary arterial shunt. Without requiring delicate manipulations of the systemic and pulmonary vascular resistances, the direct shunt constructed using a non-valved conduit of polytetraflourethylene provides not only adequate flow of blood to the lungs, but also maintains a stable systemic circulation after the first stage reconstruction.

One advantage of the direct shunt over the systemic-to-pulmonary arterial shunt is that it eliminates diastolic runoff into the pulmonary circulation with concurrent unloading of the systemic morphologically right ventricle. The resultant high diastolic pressure, and decreased volume load, may improve myocardial perfusion and ventricular function. One drawback of the shunt, however, is the volume load that occurs by reversal of the flow through the non-valved conduit. We previously reported that the greatest reversal in flow occurs within 1 month after surgery, and then decreases over time.[13] From an anatomical point of view, hearts with hypoplasia of the left ventricle and small shunts between the right ventricle and the pulmonary arteries are similar to functionally univentricular hearts with pulmonary valvar stenosis, or after banding of the pulmonary trunk. In this setting, the flow of blood to the lungs is mainly limited by the diameter of the shunt. Our results indicate that the optimal size for the shunt is 5 millimeters in patients weighing more than 2.0 kilograms, and 4 millimeters for smaller patients.

The direct shunt is particularly beneficial for small infants undergoing their initial reconstructions. The reported survival for patients weighing less than 2.5 kilograms after the Norwood procedure is still high, at between 45% and 51%.[17,18] Even the smallest polytetrafluorethylene tube, of 3 millimeters, may be too large to limit flow through the shunt in this subgroup of patients. Because of the lack of material of suitable size, a classic Blalock-Taussig shunt has been the alternative method for these infants born with a very low weight.[7] In our series, however, 8 of the 10 patients weighing less than 2.5 kilograms survived the first stage of reconstruction, with 2 of the 4 infants weighing less than 2 kilograms having a shunt constructed of 4 millimeters diameter, and the others having 5 millimeter shunts. All of the infants survived without any hemodynamic instability or right ventricular dysfunction.

The effects of a right ventriculotomy for the placement of the shunt on the function of the systemic ventricle are potentially of great concern. During follow-up in the early to medium term, nonetheless, right ventricular function as evaluated by echocardiography was acceptable, with the fractional shortening

in the 8 patients who had undergone the Fontan procedure being greater than 30%. In addition, we did not encounter ventricular arrhythmias in any of our survivors.

Deep hypothermic total circulatory arrest has been commonly used during first stage reconstructions. Even surgeons at institutions with high surgical volumes required a duration of circulatory arrest of between 44 and 57 minutes.[6,9,19] It is obvious that less experienced surgeons require longer periods of circulatory arrest, which predisposes ill infants to neurologic deficits and multi-organ failure. The strategy of continuous cerebral perfusion employed in our series minimized or avoided the use of total circulatory arrest. We have also used modified ultrafiltration following weaning from cardiopulmonary bypass to reduce the accumulation of fluids.[20] These new treatment strategies, have contributed, at least in part, to the improved outcome of initial reconstruction of patients with hypoplasia of the left heart.

We have learned several lessons during our initial five years of experience with the modified Norwood procedure using the direct shunt. First, the non-valved shunt becomes obstructive over time, particularly 3 months or more after surgery. Since we lost 2 patients early in our series from progressive obstruction in the shunt after discharge from hospital, we have checked the saturations of our patients every week at an outpatient clinic. When patients have presented with progressive desaturation, we have performed cardiac catheterization as quickly as possible. Our current strategy involves first attempting to perform balloon angioplasty for significant stenoses in either the shunt or the right and left pulmonary arteries. If angioplasty fails, or if the saturations drop again, we then proceed to perform the bi-directional Glenn anastomosis. Second, the pulmonary arteries do not grow well after placement of the shunt, as demonstrated in the patients undergoing the Fontan operation. Our results, nonetheless, indicate that the lower pulmonary vascular resistances make the subsequent Fontan procedures feasible, even in patients with small pulmonary arteries.

In conclusion, insertion of a shunt directly from the right ventricle to the pulmonary arteries during initial reconstruction provides stable hemodynamics without requiring extensive postoperative medical intervention. We believe, therefore, that improvements in the survival of infants with hypoplasia of the left heart can be accomplished by many less experienced surgeons by applying the modified Norwood procedure using the shunt placed between the right ventricle and the pulmonary arteries. The use of a technique for cardiopulmonary bypass that avoids total circulatory arrest may have a positive impact on outcome. It should be kept in mind, however, that the shunt becomes obstructive over time.

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