

# Right ventricle–pulmonary artery shunt in first-stage palliation of hypoplastic left heart syndrome

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**Objective:** Pulmonary overcirculation through a systemic-pulmonary shunt has been one of the major causes of early death after the Norwood procedure. To avoid this lethal complication, we constructed a right ventricle–pulmonary shunt in first-stage palliation of hypoplastic left heart syndrome.

**Methods:** Between February 1998 and February 2002, 19 consecutive infants, aged 6 to 57 days (median, 9 days) and weighing 1.6 to 3.9 kg (median, 3.0 kg), underwent a modified Norwood operation with the right ventricle–pulmonary artery shunt. The procedure included aortic reconstruction by direct anastomosis of the proximal main pulmonary artery and a nonvalved polytetrafluoroethylene shunt between a small right ventriculotomy and a distal stump of the main pulmonary artery. The size of the shunt used was 4 mm in 5 patients and 5 mm in 14.

**Results:** All patients were managed without any particular manipulation to control pulmonary vascular resistance. There were 17 survivors (89%), including 3 patients weighing less than 2 kg. Two late deaths occurred due to obstruction of the right ventricle–pulmonary artery shunt. Thirteen patients underwent a stage II Glenn procedure after a mean interval of 6 months, with 2 hospital deaths. To date, a stage III Fontan procedure has been completed in 4 patients. Overall survival was 62% (13/19). Right ventricular fractional shortening at the last follow-up (3–48 months after stage I) ranged from 26% to 43% (n = 13, mean, 33%).

**Conclusion:** Without delicate postoperative management to control pulmonary vascular resistance, the modified Norwood procedure using the right ventricle–pulmonary shunt provides a stable systemic circulation as well as adequate pulmonary blood flow. This novel operation may be particularly beneficial to low-birth-weight infants with hypoplastic left heart syndrome.

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**S**urvival of infants born with hypoplastic left heart syndrome (HLHS) has steadily improved since Norwood and colleagues<sup>1</sup> first reported a multistaged reconstructive approach in 1983. The Norwood procedure (stage I palliation) consists of atrial septectomy, reconstruction of the ascending aorta and aortic arch, and placement of a systemic-pulmonary shunt. 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.<sup>2,3</sup> Over the past decade, efforts to achieve a balanced circulation during the early postoperative period have focused on limiting pulmonary blood flow and improving

systemic blood flow. These measures have included reduction in the size of the systemic-pulmonary shunts,<sup>3,4</sup> ventilator manipulations according to delicate blood gas analyses,<sup>3,5</sup> use of hypoxic admixtures,<sup>6,7</sup> and systemic vasodilators.<sup>3,6,8</sup> Although several experienced centers have achieved operative survival for the Norwood procedure between 63% and 94%,<sup>3-5,9-11</sup> this procedure still remains a challenging step with high mortality for many institutions with a smaller surgical volume.<sup>12,13</sup>

To avoid hemodynamic instability associated with the systemic-pulmonary shunt, we constructed a nonvalved polytetrafluoroethylene (PTFE) shunt between the right ventricle and the pulmonary artery (RV-PA shunt) in first-stage palliation of HLHS. In this article we describe our entire experience with the RV-PA shunt, developing surgical techniques, and early- to medium-term results of this novel surgical approach.

## Patients and Methods

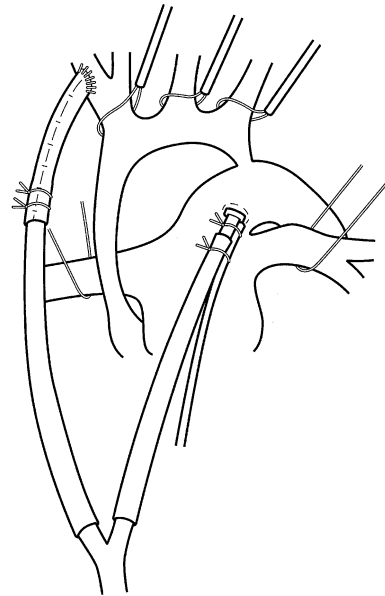
### Patient Population

Between February 1998 and February 2002, 19 consecutive infants (11 boys and 8 girls) with HLHS or a variant underwent a modified Norwood procedure using an RV-PA shunt. The diagnosis was made antenatally in 3 patients. Two were preterm infants (<37 weeks' gestational age). Three patients had ductal shock before admission. On arrival at Okayama University Hospital, all patients underwent detailed echocardiography. Fifteen patients had classic HLHS, 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 4 patients had variants of HLHS with left ventricular and aortic arch hypoplasia: 2 had a double-outlet right ventricle with aortic and subaortic stenoses, 1 had aortic and mitral stenoses with a ventricular septal defect, and 1 had aortic atresia with type B interrupted aortic arch and a ventricular septal defect. Associated anomalies included a persistent left superior vena cava in 2 patients and an aberrant right subclavian artery in 1. The diameters of the ascending aorta ranged from 1.6 to 7.6 mm (mean, 3.6 mm; median, 3.0 mm) and were 2 mm or less in 5 patients. Right ventricular fractional shortening ranged from 24% to 43% (mean, 33%; median, 32%). Tricuspid regurgitation documented by color flow Doppler echocardiography was absent in 3 patients, mild in 9, moderate in 6, and severe in 1.

Age at operation ranged from 6 to 57 days (mean, 15 days; median, 9 days). Weight at operation ranged from 1.6 to 3.9 kg (mean, 2.8 kg; median, 2.8 kg) and 3 patients weighed less than 2.0 kg. Twelve patients underwent mechanical ventilation before the operation, and all but 1 patient received an infusion of alprostadil (prostaglandin E<sub>1</sub>).

### 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. Dual arterial cannulas were inserted into the ductus arteriosus



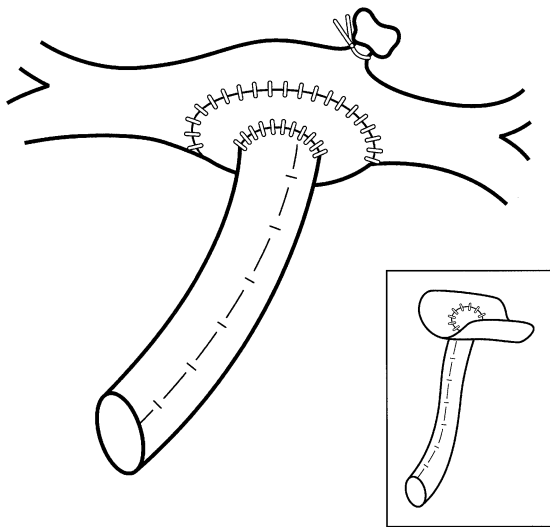
**Figure 1.** A 3-mm PTFE tube was anastomosed to the innominate artery. Dual arterial cannulas were inserted into the PTFE tube and the ductus arteriosus.

and into a 3.5-mm PTFE tube (Gore-Tex, W. L. Gore & Associates, Inc, Flagstaff, Ariz) that was anastomosed to the innominate artery. After insertion of a venous cannula into the right atrium, cardiopulmonary bypass was commenced at a flow rate of 150 to 180 mL/min per kilogram (Figure 1). The branch right and left pulmonary arteries were temporarily occluded with tourniquets and systemic cooling was initiated.

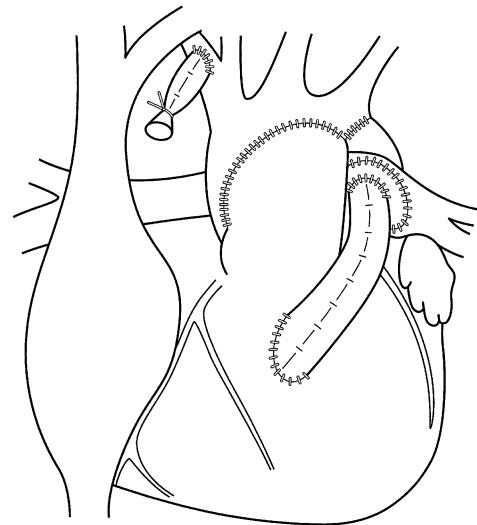
Before establishment of cardiopulmonary bypass, the shunt material was prepared on another table as follows. In case of a 5-mm shunt, a 5-mm PTFE tube was cut into an appropriate length as a shunt tube. A cuff for an anastomosis to the distal end of the main pulmonary artery was tailored by vertically 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 with a 8-0 PTFE suture (*rectangle* in Figure 2).

During the cooling phase, the isthmus was ligated and divided to obtain a better operative field. After 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 with an 8-0 PTFE suture (Figure 2). An appropriate site of ventriculotomy for the RV-PA shunt, which was 1.5 to 2 cm below the pulmonary valve, was marked on the right ventricular outflow tract. 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 6 patients, we abandoned this procedure because of intractable bleeding and the high incidence of late obstruction at the anastomotic site.

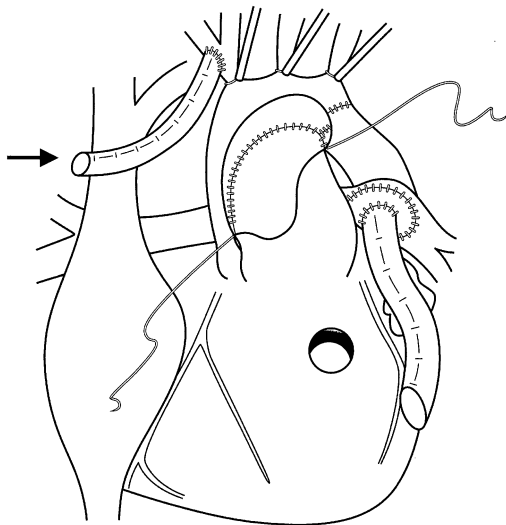
At a nasopharyngeal temperature of less than 22°C, the descending aorta was clamped. After removal of the perfusion can-



**Figure 2.** A PTFE tube was previously anastomosed to a PTFE cuff (*rectangle*). The cuff was anastomosed to the distal end of the main pulmonary artery.



**Figure 4.** After completion of aortic reconstruction and atrial septectomy, proximal anastomosis of the RV-PA shunt was performed with the heart beating.



**Figure 3.** The descending aorta was anastomosed to the posterior wall of the aortic arch. The entire aortic arch and ascending aorta were reconstructed by direct anastomosis of the proximal main pulmonary artery. A small right ventriculotomy was made in the outflow tract for proximal anastomosis of the RV-PA shunt.

nula 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.<sup>14,15</sup> 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.

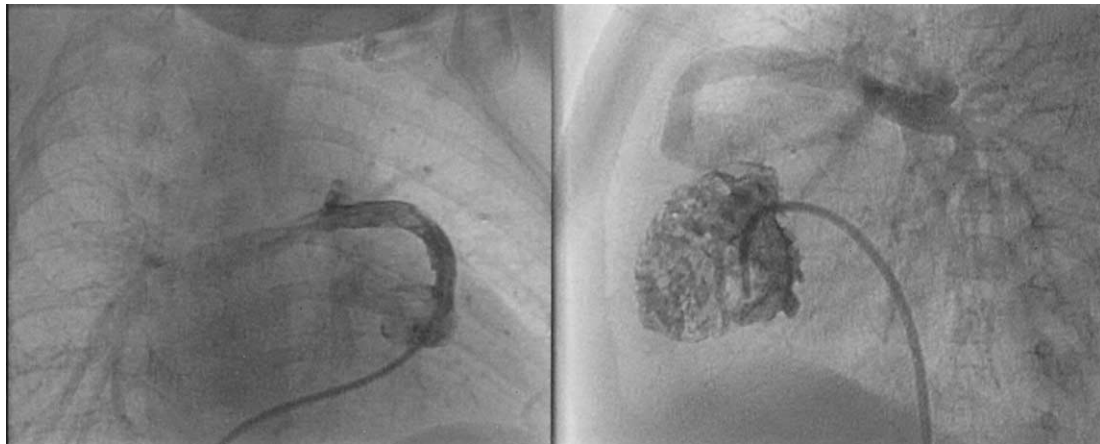
At this stage, cold crystalloid cardioplegic solution (30 mL/kg) was administered over 3 minutes either from the aortic root or

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. Cardiopulmonary bypass was resumed for isolated cerebral perfusion through the innominate artery. The aortic arch reconstruction was carried out by means of Brawn's modification.<sup>3</sup> 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 (Figure 3).

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. A small right ventriculotomy was made at the previously marked site (Figure 3). To prevent late obstruction at the shunt anastomosis, it was important to slice off a piece of ventricular muscle underlying the ventriculotomy. Cardiopulmonary bypass was reinstated, and all snares were removed. The RV-PA shunt was placed to the left of the neo-aorta in all patients and was anastomosed to the right ventriculotomy during rewarming (Figure 4). The size of the RV-PA shunt used was 4 mm in 5 patients weighing less than 2.5 kg and 5 mm in the other patients. The mean total circulatory arrest time was 6 minutes (range, 0 to 17 minutes), the mean myocardial ischemic time was 47 minutes (range, 27 to 94 minutes), and the mean descending aortic clamp time was 51 minutes (range, 10 to 99 minutes). All patients could be weaned from cardiopulmonary bypass without any difficulties.

#### Postoperative Management

Postoperative management of patients undergoing the modified Norwood operation with the RV-PA shunt was basically the same as that for neonates undergoing other types of operations. Delayed sternal closure was performed in all but 1 patient on postoperative



**Figure 5.** Frontal view at end-systole (*left*) and lateral view at end-diastole (*right*) of a right ventriculogram 4 months after stage I palliation, showing an unobstructed RV-PA shunt.

days 2 to 7 (median, 4 days). The inotropic drugs used were dopamine 5 to 10  $\mu\text{g}/\text{kg}$  per minute, epinephrine 0.1  $\mu\text{g}/\text{kg}$  per minute, and calcium chloride 0.25 mmol/h. Ventilator settings were usually constant, at a rate of 20 breaths/min, a positive end-expiratory pressure of 5 cm  $\text{H}_2\text{O}$ , and peak inspiratory pressures of 18 to 20 cm  $\text{H}_2\text{O}$ , except in case of airway problems. The inspired oxygen fraction was adjusted to keep arterial oxygen saturations higher than 75%. Systemic and pulmonary vasodilators to control vascular resistance were not used in any of the 19 patients.

#### Echocardiographic Assessment and Late Management

To evaluate diastolic reverse flow and the pressure gradient across the nonvalved RV-PA shunt, sequential Doppler echocardiography was performed in 7 patients at postoperative months 1 and 4 before stage II palliation. A spectral time-velocity display was obtained at the middle point of the RV-PA shunt. The velocity-time integrals of reverse and forward flow were measured by tracing the curves above and below the spectral baseline, respectively. The velocity-time integrals of the diastolic reverse flow were divided by the velocity-time integrals of systolic forward flow for calculation of the Doppler flow reversal ratio. Elective cardiac catheterization was carried out 3 to 4 months after stage I palliation. Results are expressed as mean  $\pm$  SD. The Wilcoxon signed-rank test was used for comparison of data between postoperative months 1 and 4.

When patients presented with progressive desaturation, the stage II bidirectional Glenn (BDG) procedure was performed with or without cardiopulmonary bypass. The RV-PA was left open as an additional pulmonary blood supply. The stage III Fontan procedure was performed at approximately 2 years of age. A lateral tunnel cavopulmonary connection was constructed by means of a PTFE patch or a right atrial flap without fenestration.

#### Results

Without any particular ventilatory manipulation, hemodynamic instability never occurred in any of the 19 patients. Transcutaneous oxygen saturations were maintained be-

tween 75% and 85%. Diastolic blood pressures remained above 40 mm Hg with pulse pressures of 20 to 30 mm Hg. There were 17 hospital survivors (89%), including 3 premature infants weighing less than 2 kg. One patient died from sudden cardiac arrest on the next day and the other from septicemia after 2 weeks. There were 2 late deaths among the first 4 patients; both died from severe hypoxemia due to obstruction of the RV-PA shunt (1 at 3 months and 1 at 4 months after surgery).

The mean Doppler flow reversal ratio in the RV-PA shunt decreased from  $0.28 \pm 0.03$  at 1 postoperative month to  $0.16 \pm 0.02$  at 4 months ( $n = 7$ ,  $P = .027$ ), while the mean pressure gradient across the shunt increased from  $34 \pm 10$  mm Hg to  $58 \pm 14$  mm Hg ( $P = .018$ ). At the same time, the mean transcutaneous oxygen saturation decreased from  $82 \pm 7\%$  to  $70 \pm 8\%$  ( $P = .018$ ). Cardiac catheterization and cineangiography before stage II palliation (Figure 5) demonstrated that the mean pulmonary artery pressure was  $11 \pm 2$  mm Hg ( $n = 13$ ; range, 9 to 15 mm Hg) and the mean PA index<sup>16</sup> measured was  $215 \pm 96$  (range, 117 to 458).

Thirteen patients underwent the BDG at a median age of 6 months (range, 3 to 9 months), and 6 of them required instrumental dilation (3-mm or 4-mm Hegar dilator) for pulmonary artery stenosis near the RV-PA shunt. There were 2 hospital deaths, one from viral pneumonia and the other from progressive hypoxemia. Currently 2 patients are awaiting the BDG, 7 are awaiting the Fontan, and 4 have undergone the Fontan, with an overall survival of 62%. The last echocardiographic evaluation was performed in 13 survivors at a median of 16 months (range, 3-48 months) after the modified Norwood procedure. Right ventricular fractional shortening ranged from 26% to 43% (mean, 33%; median, 31%). Tricuspid regurgitation was absent in 5 pa-

tients, mild in 5, and moderate in 3 whose regurgitation before stage I palliation was absent, moderate, and severe, respectively.

## Discussion

The RV-PA shunt to reestablish pulmonary blood supply in stage I palliation for HLHS was first introduced by Norwood and colleagues in 1981.<sup>17</sup> The shunt materials they used were relatively large for neonates; 8-mm nonvalved PTFE tubes in 2 patients and 12-mm valved conduits in 2. These patients all died within 11 hours after surgery either from excessive pulmonary blood flow or from right ventricular failure.<sup>2</sup> Kishimoto and coworkers<sup>18</sup> in Japan revived the RV-PA shunt using a xenopericardial valved conduit, and they reported stable postoperative hemodynamics with a high diastolic blood pressure. Their findings stimulated us to attempt an RV-PA shunt, but with a small nonvalved PTFE conduit in stage I palliation. Although our institutional results of reconstructive surgery for HLHS before 1998 included a survival of 53% (17/33 classic Norwood over 6 years), we could achieve a stage I survival of 89% with the application of the RV-PA shunt and subsequently reach an overall survival of 62%.

The results of the present study have clearly shown differences in postoperative hemodynamics between the systemic-pulmonary shunt and the RV-PA shunt. Without delicate manipulation of systemic and pulmonary vascular resistance, the RV-PA shunt using a nonvalved PTFE conduit provides a stable systemic circulation as well as adequate pulmonary blood flow after stage I palliation. Although we have not determined an optimal size for the RV-PA shunt, our current choice of shunt is a 5-mm PTFE tube for patients weighing more than 2 kg and a 4-mm tube for patients less than 2 kg. These sizes of shunts are almost equivalent to 50% of the predicted normal size of the main pulmonary artery.<sup>19</sup> From an anatomic point of view, therefore, HLHS hearts after aortic reconstruction with the main pulmonary artery and placement of an RV-PA shunt are similar to univentricular hearts with pulmonary stenosis. In this anatomic setting, pulmonary blood flow is limited by the size of the shunt, and systemic circulation, including coronary perfusion, theoretically cannot be compromised by changes in pulmonary vascular resistance. Our hypothesis was confirmed in other type of single-ventricle physiology as well. We have applied the RV-PA shunt in 2 newborn infants with heterotaxy syndrome, pulmonary atresia, and infracardiac type total anomalous pulmonary venous connection. Postoperative hemodynamics of these patients was also stable and both survived.

Excellent hemodynamics provided by the RV-PA shunt is particularly beneficial for low-birth-weight infants undergoing stage I palliation. Reported survival for

patients weighing less than 2.5 kg after the Norwood procedure is still high (45% to 51%).<sup>20,21</sup> Even the smallest 3-mm PTFE tube may be too large to limit pulmonary blood flow through the systemic-pulmonary shunt in this subgroup of patients. Because of the lack of suitably sized material, a standard Blalock-Taussig shunt has been the alternative method for very-low-weight infants.<sup>4,22</sup> In the present series, the 3 infants weighing less than 2 kg received a 4-mm RV-PA shunt and all survived without any hemodynamic instability or right ventricular dysfunction.

The effects of a right ventriculotomy for the placement of the RV-PA shunt on systemic right ventricular function are of great concern. During early- to medium-term follow-up, right ventricular function evaluated by echocardiography was acceptable, and its fractional shortening in the 4 patients who had undergone the Fontan procedure was higher than 30%. In addition, ventricular arrhythmias were not found in any of the survivors. Another concern is related to the degree of flow reversal in the nonvalved RV-PA shunt. Doppler echocardiography demonstrated the greatest reversal flow ratio within 1 month after surgery, but all patients tolerated this volume overload well, maintaining reasonable oxygen saturation levels.

The lessons learned from 4 years of experience with the modified Norwood procedure using the RV-PA shunt are twofold. First, the nonvalved PTFE RV-PA shunt becomes obstructive with time, particularly from 3 months after surgery, as evidenced by the decrease in oxygen saturation and increase in the pressure gradient across the shunt. As 2 patients in the early series died from progressive shunt obstruction after hospital discharge, we are now very careful when patients' oxygen saturations start decreasing. We have not used a larger shunt, such as 6-mm or 8-mm PTFE tube, because effect of a larger ventriculotomy on ventricular function is unclear. Second, pulmonary artery growth in patients who received the RV-PA shunt is not as well as in those who had a systemic-to-pulmonary shunt, indicating that the RV-PA shunt provides less pulmonary blood flow than the systemic-to-pulmonary shunt. However, resultant lower pulmonary vascular resistance made the BDG and subsequent Fontan procedures possible, even in patients with a PA-index of 200 or less.

In conclusion, the RV-PA shunt in stage I palliation provides stable hemodynamics and fine control of systemic and pulmonary resistance without extensive postoperative medical intervention. This novel procedure may be particularly beneficial to low-birth-weight infants with HLHS. However, it is cautioned that the RV-PA shunt becomes obstructive with time. We believe that improvement in survival for infants undergoing stage I palliation for HLHS would be reproducible for many less experienced surgeons with application of the RV-PA shunt.

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## Discussion

**Dr S. Bert Litwin** (*Milwaukee, Wis*). Dr Sano and his colleagues have presented data on 19 infants who underwent Norwood palliation for HLHS, and they achieved an excellent 89% hospital survival. Their surgical technique differs from that used by most surgeons in the current era with pulmonary blood flow received from a direct RV-PA shunt rather than a systemic-pulmonary artery shunt.

Placing the pulmonary and systemic circulations in parallel rather than in series may have certain advantages. Imbalance of pulmonary and systemic blood flows is the leading cause of morbidity and mortality after the Norwood procedure. Most centers accomplish balance by manipulating vascular resistance. Dr Sano's patients required no such manipulation, which resulted in a more predictable postoperative recovery.

The pulmonary bed was not subjected to nor dependent on diastolic flow, and there should be less change in pulmonary blood flow with pulmonary hypertensive crises or during resuscitation in the presence of low cardiac output or after a cardiac arrest.

There may, however, be deleterious effects with an additional volume load on the ventricle from regurgitation through the RV-PA shunt. This may result in ventricular dilatation and tricuspid regurgitation. A number of patients presented here had tricuspid regurgitation after stage I, although the incidence was less than preoperatively.

Dr Sano used larger 4- and 5-mm shunts and achieved balanced pulmonary and systemic circulations without overperfusion of the lungs. With larger shunts, less precision in gauging the size of the shunt should be needed and this might avoid some early reoperations for shunt change. Also, with a larger shunt, there should be a greater shunt longevity as well as a slower and more predictable shunt closure pattern. Such was not the case here in that 2 patients died late due to acute shunt closure and 7 others studied by echocardiography showed progressive shunt narrowing within 4 months.

I have a few questions.

Did the shunt regurgitation cause an increase in tricuspid regurgitation in any patient and did you have to repair the tricuspid valve at stage II in any patient?

Why did you leave the shunt patent, as stated in your manuscript, after stage II? Were the 2 deaths after stage II related to residual shunt patency or tricuspid regurgitation? Did any patients after stage II have prolonged pleural drainage?

In the manuscript you state that your survival was 53% before you used the RV-PA shunt and 89% with the RV-PA shunt. Did you analyze the two groups for other factors that might have accounted for your improved survival?

Dr Sano, your presentation was clear, your manuscript was well written, and you have made a compelling argument for the use of the RV-PA shunt in the Norwood procedure. This information should be useful to surgeons in considering various options for babies with HLHS, and I thank you for bringing this information to this meeting.

**Dr Sano.** Thank you, Dr Litwin, for your comments.

Regarding your first question, we also were concerned about volume loading due to shunt regurgitation through a nonvalved conduit. This was also why we used a 4-mm PTFE graft rather than a 5-mm graft initially.