**Background:** Significant tricuspid valve regurgitation (TR) is considered a poor prognosis factor for patients with hypoplastic left heart syndrome (HLHS). Performing a tricuspid valve plasty (TVP) during Norwood Stage I palliation (S1P) has rarely been reported. We report mid-term results of performing TVP during Norwood S1P.

**Methods and Results:** Between December 2004 and July 2013, 48 patients with HLHS or variants underwent Norwood S1P and of them 11 (23%) with TR of a moderate degree or above underwent TVP. The estimated 1- and 5-year survival rates for Norwood S1P were 61% and 54%, respectively. Among the 11 patients with TVP, there were 2 early deaths. Nine patients had a Stage II operation and 3 of them died late. Four patients completed a Fontan operation, and 2 were waiting. Using Cox regression analysis, lower body weight, presence of intact atrial septum, and preoperative cardiopulmonary resuscitation were factors associated with increased risk for death. Patients with significant TR and undergoing TVP during Norwood S1P had a similar survival curve to those without significant TR.

**Conclusions:** Our results for TVP performed during Norwood S1P were encouraging. They suggested that aggressive TVP is warranted in Norwood S1P when primary heart transplantation is not available. Further study is required to determine if the strategy does improve the results for patients with HLHS and TR initially.  

**Key Words:** Hypoplastic left heart syndrome; Norwood Stage I palliation; Tricuspid valve plasty; Tricuspid valve regurgitation

Significant tricuspid valve regurgitation (TR) is considered a poor prognosis factor for patients with hypoplastic left heart syndrome (HLHS).^1\textsuperscript{-3} Barber et al showed that without intervention for moderate or severe TR, most patients had no significant change in the degree of TR after Norwood staged reconstruction and a significantly worse survival (survival, 55% vs. 35% at 1 year after Norwood procedure).^4\textsuperscript{-}Infant heart transplantation is advocated for HLHS with significant TR, but infant heart transplantation for HLHS is not feasible in Taiwan because of the lack of donors. Similar to Japan, where heart transplantation is very limited, a Japanese series also showed significant TR associated with higher mortality rates.^2\textsuperscript{-} Several reports have shown that tricuspid valve plasty (TVP) in HLHS patients has a positive effect on survival and right ventricular function.^10\textsuperscript{-}13 However, in most cases the TVP was preformed after Norwood S1P (mainly during Stage II operation). Performing a TVP during Norwood S1P is rarely reported. Because of the patients’ limited chances of survival, we have adopted an aggressive policy of TVP performed during Norwood S1P and we report our mid-term results.

**Methods**

We performed a single-center retrospective study of 48 consecutive infants at the National Taiwan University Hospital who all underwent a Norwood S1P for HLHS or its variants between 2004 and 2013.

Institutional ethics board approval was obtained, and the medical and surgical records, as well as the angiographic and echocardiographic data for all patients were reviewed. Two patients underwent a hybrid approach (bilaterial PA banding and ductal stenting) initially, but both were converted to...
Techniques Used for Tricuspid Valve Repair (TVP)

We used a saline-filled 20-ml syringe with a 14-gauge catheter to test the tricuspid valve. During Norwood S1P, the TVP was primarily an annuloplasty or commissuroplasty procedure, because the leaflet itself is too fragile to hold stitches. Initially, a De Vega-type annuloplasty over the anterior-posterior leaflets was performed (n=8). The target annulus diameter was about the normal tricuspid valve size of the neonate (∼13 mm in diameter). Recently, we performed posterior leaflet obliteration (n=3). We also try to find the specific leaking commissure and apply a 5-0 pledget prolene suture accordingly. The primary goal at this stage is to improve coaptation of the TV leaflets and commissures. In TVP performed at a later stage (bidirectional Glenn (BDG) shunt or Fontan operation), an edge-to-edge suture technique for improving the coaptation was also used in addition to the annuloplasty/commissural plication techniques.

Statistical Analysis

The primary outcomes of this study were death after Norwood S1P and the all-cause mortality rate during the follow-up period. Norwood S1P mortality rate was defined as any deaths before a Stage II operation, including all-cause hospital and inter-stage mortality rates.

Statistical analysis was performed using R 3.0.2 software (R Foundation for Statistical Computing, Vienna, Austria). In statistical testing, a 2-sided P value ≤0.05 was considered statistically significant. The distributional properties of continuous variables are expressed by mean±standard deviation.
analysis was performed by an expert biostatistician. The statistical significances between the forward and backward steps) was applied to finally, the stepwise variable selection procedure (with iterations) was used in our regression analyses. Specifically, basic model-fitting techniques for (1) variable selection, (2) goodness-of-fit assessment, and (3) regression diagnostics were used in our regression analyses. Specifically, the stepwise variable selection procedure (with iterations between the forward and backward steps) was applied to obtain the best candidate final regression model. The statistical analysis was performed by an expert biostatistician.

### Results

#### Demographic Data
There were 31 (64.6%) male and 17 female patients. The Norwood S1P was performed at a median age of 3.5 [1–9.5] days and median body weight of 3.0 [2.7–3.3] kg. The median CPB time was 207 [181–258] min, and the median low-flow duration was 63 [42.5–74.5] min. There were 15 (31.3%) patients diagnosed prenatally. Before the Norwood operation, shock or intubation was noted in 32 (67%), and cardiac arrest requiring CPR was noted in 4 (8.3%) of them. Classical HLHS was present in 32 (67%) and aortic atresia was present in 22 (46%).

#### Summary of Patients’ Outcomes
The patients’ outcomes are summarized in Figure 1. Of the 48 patients, 13 died before BDG shunt was performed; 32 patients underwent a BDG as a second-stage operation. Between the BDG and total cavopulmonary connection (TCPC), 5 patients died early (all related to heart failure and TR) and 3 died late. Finally, 14 patients underwent TCPC, and 10 patients were waiting for TCPC.

### Patients With TVP Performed During Norwood S1P
In the cohort, there were 11 (23%) patients who underwent TVP during Norwood S1P; moderate, moderate to severe, and severe TR were noted in 3, 3, and 4 patients, respectively. The details of this group are summarized in Table 1. Intraoperatively, most patients were considered to have tricuspid annular dilation, and poor coaptation of the anterior-septal leaflet or posterior-septal leaflet was also a prominent finding. Annular plication with either a De Vega-type suture (n=8), or more focus on the posterior leaflet similar to posterior leaflet obliteration (n=3) was performed. Two patients died in the early posterior period: 1 patient had suspected kinking of the

### Table 1. Summary of 11 Hypoplastic Left Heart Syndrome Patients With TVP Performed During Norwood S1P

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Diagnosis</th>
<th>Body weight (kg)</th>
<th>Operative age (days)</th>
<th>TR degree</th>
<th>Operative findings</th>
<th>TVP technique</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>AA, MS</td>
<td>3.3</td>
<td>2</td>
<td>Moderate</td>
<td>Tricuspid valve septal leaflet cleft</td>
<td>Cleft closure with De Vega+obliterate mitral valve</td>
<td>Complete TCPC (mild TR)</td>
</tr>
<tr>
<td>2</td>
<td>AA, MA</td>
<td>2.2</td>
<td>3</td>
<td>Moderate</td>
<td>Annular dilation, posterior-septal commissure leakage</td>
<td>De Vega</td>
<td>Death (shunt kinking)</td>
</tr>
<tr>
<td>3</td>
<td>AA, MA, IAS</td>
<td>2.8</td>
<td>0</td>
<td>Severe, poor RV contractility</td>
<td>Central jet</td>
<td>De Vega</td>
<td>Death (hypoxemia)</td>
</tr>
<tr>
<td>4</td>
<td>AA, MA</td>
<td>3.5</td>
<td>8</td>
<td>Moderate</td>
<td>Central jet, with leaflet tethering</td>
<td>De Vega</td>
<td>Complete TCPC (trivial TR)</td>
</tr>
<tr>
<td>5</td>
<td>AS, MS</td>
<td>3</td>
<td>31</td>
<td>Severe</td>
<td>Multiple leakage sites</td>
<td>De Vega</td>
<td>Death after BDG with second TVP (MRSA sepsis)</td>
</tr>
<tr>
<td>6</td>
<td>AS, MS, preOp CPR</td>
<td>3.5</td>
<td>1</td>
<td>Severe</td>
<td>Poor coaptation of posterior-septal commissure</td>
<td>5–0 prolene pledget on posterior leaflet</td>
<td>Death after BDG (renal failure)</td>
</tr>
<tr>
<td>7</td>
<td>AA, MA</td>
<td>2.5</td>
<td>5</td>
<td>Moderate-severe</td>
<td>Central jet dominantly</td>
<td>De Vega</td>
<td>Re-do TVP at BDG, complete TCPC (mild TR)</td>
</tr>
<tr>
<td>8</td>
<td>AA, MA</td>
<td>3.7</td>
<td>11</td>
<td>Moderate-severe</td>
<td>Tricuspid valve tethering, central jet and multiple jets due to poor coaptation zone</td>
<td>De Vega</td>
<td>Re-current TR, CPR,ECMO, Death after BDG</td>
</tr>
<tr>
<td>9</td>
<td>AA, MA</td>
<td>3.2</td>
<td>3</td>
<td>Severe</td>
<td>Central jet, septal leaflet tethering</td>
<td>De Vega</td>
<td>Complete TCPC (mod TR, TVP at TCPC)</td>
</tr>
<tr>
<td>10</td>
<td>AA, MA</td>
<td>3</td>
<td>9</td>
<td>Moderate-severe</td>
<td>Central jet; annular dilation, leaflet tethering</td>
<td>5–0 prolene pledget on post-septal commissure</td>
<td>Wait for TCPC</td>
</tr>
<tr>
<td>11</td>
<td>AA, MA</td>
<td>3.6</td>
<td>2</td>
<td>Moderate-severe</td>
<td>Posterior-septal commissure downward displacement; RV dysfunction</td>
<td>5–0 prolene pledget on post-septal commissure</td>
<td>Wait for TCPC (moderate TR)</td>
</tr>
</tbody>
</table>

AA, aortic atresia; AS, aortic stenosis; BDG, bidirectional Glenn shunt; IAS, intact atrial septum; MA, mitral atresia; MS, mitral stenosis; RV, right ventricle; TCPC, total cavopulmonary connection; TR, tricuspid regurgitation; TVP, tricuspid valve plasty; De Vega, De Vega's annuloplasty (5–0 prolene pledget).
TVP in Norwood S1P

Table 2. Risk Factors for Overall Mortality Before Stage II Operation in HLHS Patients

<table>
<thead>
<tr>
<th></th>
<th>Survivors (n=35)</th>
<th>Non-survivors (n=13)</th>
<th>Univariate P value</th>
<th>Multivariate P value</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>M/F</strong></td>
<td>22/13</td>
<td>9/4</td>
<td>0.747</td>
<td>0.0027</td>
</tr>
<tr>
<td>Body weight (kg)</td>
<td>3.2±0.4</td>
<td>2.6±0.6</td>
<td>0.236</td>
<td>0.000023–0.006</td>
</tr>
<tr>
<td>&lt;2.5kg (%)</td>
<td>2 (5.7%)</td>
<td>6 (46%)</td>
<td>0.003</td>
<td></td>
</tr>
<tr>
<td>Prenatal diagnosis (%)</td>
<td>12 (34.3%)</td>
<td>3 (23%)</td>
<td>0.727</td>
<td></td>
</tr>
<tr>
<td>PreOp shock/intubation (%)</td>
<td>22 (62.9%)</td>
<td>10 (83.3%)</td>
<td>0.49</td>
<td></td>
</tr>
<tr>
<td>PreOp CPR (%)</td>
<td>2 (5.7%)</td>
<td>2 (15.4%)</td>
<td>0.294</td>
<td>60.2</td>
</tr>
<tr>
<td>Classical HLHS (%)</td>
<td>25 (71.4%)</td>
<td>7 (53.8%)</td>
<td>0.31</td>
<td>2.68–3.803</td>
</tr>
<tr>
<td>Associated anomaly (%)</td>
<td>2 (5.7%)</td>
<td>2 (15.4%)</td>
<td>0.29</td>
<td></td>
</tr>
<tr>
<td>Aortic atresia (%)</td>
<td>15 (42.9%)</td>
<td>7 (53.8%)</td>
<td>0.53</td>
<td></td>
</tr>
<tr>
<td>AsAo diameter (mm)</td>
<td>3.3±1.4</td>
<td>2.9±1.3</td>
<td>0.599</td>
<td></td>
</tr>
<tr>
<td>IAS (%)</td>
<td>1 (2.9%)</td>
<td>4 (30.8%)</td>
<td>0.015</td>
<td>5,288.3</td>
</tr>
<tr>
<td>Significant TR (%)</td>
<td>10 (28.6%)</td>
<td>2 (15.4%)</td>
<td>0.46</td>
<td>44.1–5,721,265</td>
</tr>
<tr>
<td>TVP (%)</td>
<td>9 (25.7%)</td>
<td>2 (15.4%)</td>
<td>0.702</td>
<td></td>
</tr>
<tr>
<td>BT shunt/RV-PA shunt</td>
<td>4/31</td>
<td>3/10</td>
<td>0.37</td>
<td></td>
</tr>
<tr>
<td>Low flow/DHCA (min)</td>
<td>59±30</td>
<td>62±20</td>
<td>0.112</td>
<td></td>
</tr>
<tr>
<td>Cross-clamp (min)</td>
<td>97±27</td>
<td>95±24</td>
<td>0.771</td>
<td></td>
</tr>
<tr>
<td>CPB duration (min)</td>
<td>208±54</td>
<td>265±77</td>
<td>0.094</td>
<td></td>
</tr>
</tbody>
</table>

AsAo, ascending aorta; BT shunt, modified Blalock-Tausig Shunt; CI, confidence interval; CPB, cardiopulmonary bypass; CPR, cardiopulmonary resuscitation; DHCA, deep hypothermia circulatory arrest; HLHS, hypoplastic left heart syndrome; OR, odds ratio; PreOp, preoperative; RV-PA, right ventricle to pulmonary artery; TR, tricuspid valve regurgitation. Other abbreviations as in Table 1.

RV-PA shunt, resulting in sudden cardiac arrest, and died despite ECMO support and revision of the shunt. The other patient had an IAS and underwent Norwood S1P soon after birth but still had persistent hypoxemia after the operation and died.

Among the survivors (n=9), moderate residual TR was noted in only 1 patient, and the other patients had trivial (n=2) or mild (n=6) residual TR.

Before Stage II (BDG shunt), 1 patient developed severe TR and received ECMO support during CPR (ECPR). The patient underwent TVP again and ECMO could be weaned-off postoperatively. The previous annuloplasty suture (5-0 prolene) was found to have detached from the annulus, resulting in severe TR. However, the patient died of persisted heart failure after the Stage II operation.

The other 8 patients successfully survived to Stage II operation, during which 2 patients underwent TVP again. The edge-to-edge suture (Alfieri stitches) technique was used in 1, and De Vega annuloplasty with 1 edge-to-edge suture were used in the other patient. Both patients’ TR became trivial after the operation.

Between Stages II and III, 2 patients died; 1 from staphylococcal bacteremia, and the other died of renal failure for which the family refused further treatment.

In all, 4 patients successfully underwent TCPC as a Stage III operation. During the operation, 1 patient had TVP again for moderate TR. Posterior leaflet obliteration was performed (previous operation was De Vega’s annuloplasty). All patients survived TCPC. The other 2 patients were waiting for TCPC completion. Of the 6 survivors, all had trivial or mild TR at the last echocardiographic follow-up.

In summary, among 11 patients, 4 (36%) required additional TVP: 1 before Stage II, 2 at Stage II, 1 at Stage III; 2 of the 4 patients died before completion of TCPC.

Patients Without TVP Performed During Stage I

A total of 37 patients underwent Norwood S1P without TVP. All patients had trivial or mild TR, except 1 patient with moderate TR before operation. There were 11 (30%) deaths before Stage II operation, so 23 underwent BDG shunt as the Stage II operation, and 2 patients underwent conduit change while considering between a Fontan pathway or possible late biventricular repair. One patient is waiting for the Stage II operation. After BDG shunt operation, 2 patients died early and 3 died late. One of the patients who died early had significant TR and underwent Norwood S1P and BDG shunt with the hope of spontaneous recovery of TR, but the procedure failed. The other patient who died early underwent BDG shunt plus TVP, as described next. All 3 patients who died late lived relatively far from the hospital and they died out of the hospital, so the exact causes of the deaths were not clear. There were 10 patients who underwent TCPC successfully, and 8 patients were waiting for TCPC.

Four patients within the group without TVP during Norwood S1P underwent TVP after Norwood S1P. One patient had HLHS with pulmonary valve stenosis and had TVP before the Stage II operation. The patient successfully survived BDG shunt and is now waiting for Stage III operation (this case was previously reported). Two patients had TVP at Stage II operation. One patient had HLHS and partial anomalous pulmonary venous connection. The patient experienced heart failure, and severe TR was noted. Despite BDG shunt and TVP, the patient died as a result of persistent heart failure after operation. The other one with moderate TR and residual coarctation of aorta underwent aortoplasty, TVP and BDG shunt successfully. The patient completed TCPC operation and has been doing well to date.

One additional patient had TVP at TCPC operation. The patient underwent posterior leaflet obliteration because mild to moderate TR was noted by transesophageal echocardiography during operation. The patient had only trivial TR 3 years after operation.
**Table 3.** Cox Regression Analysis for Risk Factors for All-Cause Mortality in Hypoplastic Left Heart Syndrome Patients

<table>
<thead>
<tr>
<th>Risk Factor</th>
<th>HR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Body weight (kg)</td>
<td>0.087</td>
<td>0.027–0.280</td>
</tr>
<tr>
<td>Intact atrial septum</td>
<td>31.3</td>
<td>6.27–156.5</td>
</tr>
<tr>
<td>Preoperative cardiopulmonary resuscitation</td>
<td>7.8</td>
<td>1.84–33.3</td>
</tr>
</tbody>
</table>

CI, confidence interval; HR, hazard ratio.

**Figure 2.** Kaplan-Meier estimate of survival of (A) the whole cohort, with 95% confidence interval and (B) patients with and without tricuspid valve plasty (TVP) during Norwood Stage I palliation. Both groups have similar survival curves. Norwood S1P, Norwood Stage I palliation.
Survival Analysis

Factors for Norwood S1P Mortality For any deaths (n=13) before Stage II operation, univariate analysis showed that patients with low body weight (<2.5 kg) and IAS were significantly associated with death (Table 2). Prenatal diagnosis, associated anomaly, presence of aortic atresia, and significant TR were not significantly different between survivors and non-survivors. The duration of CPB, aorta cross-clamping, low flow and deep hypothermic circulatory arrest were not significantly different between the 2 groups.

In this cohort, IAS was associated with 80% (4/5) mortality rate and body weight <2.5 kg was associated with 75% (6/8) mortality rate. Cardiac arrest and CPR were performed in 4 patients before Norwood S1P, and 2 (50%) of them died of multi-organ failure. These 3 factors accounted for the majority (12/13) of deaths after Norwood S1P. In the multivariate analysis, these same 3 factors were significantly associated with death before Stage II palliation (Table 3).

Cox Regression Analysis The Kaplan-Meier estimate of survival is plotted in Figure 2A. The estimated 1- and 5-year survival rates were 61% and 54%, respectively. The patients with low body weight (≤2.5 kg) and IAS were associated with
Discussion

Improving the outcome of patients with HLHS or its variants remains a challenge for pediatric cardiac surgeons. In this series, we demonstrated that HLHS with TR still could be considered for Norwood staged reconstruction with aggressive TVP, and survival was similar to those without TR initially in the mid-term follow-up.

Risk Factors for Norwood S1P Mortality

Low body weight was significantly associated with higher mortality, which is similar to the literature.\(^2\)\(^–\)\(^4\) The presence of an intact or highly restricted atrial septum was also associated with higher mortality for Norwood S1P because of poor lung function.\(^5\)\(^–\)\(^7\) The patients with preoperative shock also had poor outcome after Norwood S1P.\(^8\)\(^–\)\(^10\) but in this series, preoperative CPR instead of shock was associated with significantly increased mortality. We think these 3 groups of patients would require an alternative strategy such as bilateral PA banding and an open inter-atrial septum instead of going directly to Norwood S1P.\(^11\)\(^–\)\(^13\) Without these 3 factors, survival would be much better.

TR is an important factor for the death of patients with HLHS.\(^1\)\(^–\)\(^3\)\(^,\)\(^9\)\(^,\)\(^13\)\(^–\)\(^15\) In the present cohort, TVP was performed in 23% of the patients during S1P, and the incidence of significant TR was similar to reported studies from Texas (21% before S1P).\(^1\) In contrast to other groups, where in most cases TVP was performed with BDG shunt,\(^8\)\(^–\)\(^10\)\(^,\)\(^13\)\(^,\)\(^26\) we adopted a policy of early repair of significant TR concomitantly during Norwood S1P. The reason was our early experience of losing several patients with persistent TR after Norwood S1P. Patients with pre-S1P TR might be assigned to neonatal heart transplant in North America,\(^9\) but because heart transplantation is not a feasible option in Taiwan, we believe trying to repair TR might have a positive effect on the patient’s survival.

Rational for TVP and Repair Technique

The mechanism of TR in HLHS is multi-factorial. The 3 major factors that trigger the development of significant TR are volume overload, structural tricuspid valve abnormalities, and myocardial damage/ischemia.\(^6\) In addition, residual coarctation of aorta after Stage I repair must also be excluded.

Effect of Volume Overload

The patients with HLHS before Norwood S1P and after Norwood S1P were all in-parallel circulation. The right ventricle is subject to volume overload until the BDG shunt operation, when the volume was unloaded. At this stage, TR might improve in some patients.\(^13\)\(^–\)\(^29\) The recent report by Munich’s group showed that despite ventricular unloading, the grade of TR did not improve.\(^30\) The mean Z value of the tricuspid annulus stayed the same in patients with significant TR (grade III or IV) after bidirectional superior cavopulmonary anastomosis but decreased in the patients without significant TR. Taking this into consideration, it might be reasonable to perform the annuloplasty procedure in patients with significant TR instead of expecting an improvement with volume unloading only.

Tricuspid Valve Abnormality

Structural TV abnormality remains a difficult problem. Theoretically, specific techniques for specific lesions is the preferred approach to repairing a valve, such as mitral valve plasty. Dysplastic leaflets, tethered, or prolapsed leaflets with reduced coaptation can also contribute to significant valve regurgitation. Other abnormalities included bileaflet or quadricleaflet TV, accessory orifices, prolapse of the anterior leaflet, and, rarely, Ebstein’s anomaly had been reported.\(^5\) In this condition, in addition to annuloplasty, various techniques, including repair of cleft leaflets, edge-to-edge repair of the superior and septal leaflets to primary commissural closure, and, less frequently, mobilization of the septal leaflet and chordal shortening could be considered.\(^12\)\(^,\)\(^27\)\(^–\)\(^31\)

However, repairing the TV in HLHS neonates is jeopardized by the small, fragile leaflet, and the duration of myocardial ischemia. We primarily only sutured the annulus, instead of leaflets or subvalvular tissue, in neonatal valve repair in a single ventricle. Furthermore, we gradually shifted from a non-specific, circular De Vega type annuloplasty to specific commissuroplasty such as posterior leaflet obliteration, because that technique might better address the specific leaking commissure.\(^5\)\(^,\)\(^12\) This procedure did improve TR grade in the majority of patients and did not require much more myocardial ischemia time to perform during Norwood S1P operation. A similar principle was also advocated by Sakamoto et al for repairing atrioventricular valves in patients with a single ventricle.\(^32\) If patients require TVP in the later stage (BDG shunt or TCPC), we performed more leaflet repair in addition to the annulomissuroplasty technique.

Myocardial Damage/Ischemia

HLHS patients, especially with aortic atresia, are at risk of myocardial ischemia. Preoperative shock, intraoperative myocardial ischemia, and postoperative shock all could contribute to myocardial damage and result in TR and RV dysfunction. The literature shows that patients with preserved RV function have good survival after successful TVP.\(^8\)\(^–\)\(^11\) If RV function is very poor, performing TVP might not be able to save the patient’s life.

Results of TV Repair

The results of TVP during Norwood S1P are not clear yet. In most reported series, TVP was performed in survivors of Norwood S1P. Elmi et al reported that 25% of survivors underwent TV intervention after Norwood S1P, and the 5-year survival was 82% after intervention.\(^10\) Ohye et al\(^11\) reported 27 patients with TVP after Norwood S1P: 17 (63%) patients had a successful result and 10 (37%) had an adverse outcome. Survival was 94% (16/17) for patients with a successful late result vs. 20% (2/10) for those with a poor outcome.

Nakata et al reported 16 cases of neonatal or infant atrioventricular valve repair for single-ventricle palliation, 5 of them with Norwood S1P. The exact survival was not unclear, but the diagnosis of HLHS was significantly associated with worse outcome, and overall 1-year survival was approximately 40% following valve repair.\(^32\)

Honjo et al\(^3\) reported a series of 57 patients with atrioventricular valve plasty for single-ventricle physiology; 67% of the valves were tricuspid. Survival in the repair group was lower than in the matched controls (78.9% vs. 92.7% at 1 year; 68.7% vs. 90.6% at 3 years, P=0.015). Patients with successful repair and normal ventricular function had equivalent survival to matched controls.

In the present series, although limited by case numbers (n=11) and duration of follow-up, the survival curve was similar to that for patients without TVP during Norwood S1P.
Although the mortality rate remained high (58% 1-year survival rate), the results appear encouraging for patients with an extremely poor prognosis.

**TVP at the Later Stage**

Among the 35 Norwood S1P survivors, 8 (23%) of them required additional TVP (36% (4/11) for those with initial TVP and 11% (4/37) for those without initial TVP). This incidence is similar to another reported series.\(^6\) We think the initial TVP by annulus/commissuroplasty technique had a reasonable success rate, but one-quarter of the patients may have also had other mechanism of TR, thus requiring additional repair technique. The patients without initial TR might also develop significant TR after Norwood S1P, and the residual coarctation, intrinsic TV abnormality, and annular dilatation should be addressed during the staged reconstruction strategy. Maintaining RV function remains the most important and difficult issue for patients with a single, functional RV.

**Bilateral PA Banding as Initial Approach**

Bilateral PA banding with ductal stenting or maintained by PGE1 infusion is an alternative approach to Norwood S1P.\(^9\) Guleserian et al\(^2\) reported bPAB for high-risk single-ventricle patients (n=24); among them, 7 had moderate or greater atriovenricular valve regurgitation. In the present report, patients with severe TR were contraindicated for Norwood S1P and bridge to primary cardiac transplantation. Gomide et al reported a series using bilateral PA banding to resuscitate high-risk patients, but only 1 patient had severe TR and died after Norwood S1P.\(^3\) Sugirua et al reported their experience of bilateral PA banding (n=9) for early significant TR in HLHS, and of their patients 3 died before Norwood-Glenn operation.\(^3\)

We think bilateral PA banding is beneficial for patients with pulmonary overflow, but not helpful for other TR mechanisms such as coarctation, TV abnormality, or coronary ischemia from a small ascending aorta. Because of the limited information, how to select the most appropriate approach for specific patients requires more studies.

**Study Limitations**

This study is limited by the case numbers and as a single-center study. The echocardiography was not performed by single physician, so inter-observer variation might exist. Accumulation of experience, surgical technique and management might evolve over time. Variation in case selection could have had an effect on the results. The development of a hybrid technique for HLHS might also change the management strategy. Additional study and more cases are required before it could be generalized to other centers or patients.

**Conclusions**

The results of TVP performed during Norwood S1P were encouraging. The results suggested that such an aggressive policy is warranted when primary heart transplantation is not available. Further study is required to determine if the strategy does improve the results for patients with HLHS and TR initially.

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**Conflict of Interest**

The authors did not have conflicts of interest related to this manuscript.

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