The Norwood procedure for hypoplastic left heart syndrome: results of the King Chulalongkorn Memorial Hospital

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**Background:** The Norwood procedure is often performed to treat hypoplastic left heart syndrome (HLHS). Only Blalock-Taussig (BT) shunt is used with the Norwood procedure in our institute.

**Objective:** To analyze the first-stage palliation for HLHS.

**Patients and methods:** The Norwood procedure with right modified BT shunt was performed in 26 patients with HLHS between August 1996 and November 2008. The first four patients were performed using only autologous great vessel tissue (group 1), and the other 22 patients were operated with the Norwood procedure using a homograft for arch reconstruction (group 2).

**Results:** The hospital mortality was 50.0% in group 1 and 18.2% in group 2. The overall hospital mortality was 23.1%. Four out of twenty survivors (20%) had the modified Fontan procedure.

**Conclusion:** Only a limited number of pediatric cardiac centers offered surgical treatment of the hypoplastic left heart syndrome. The survival rate in our study was in acceptable range despite the limited resource.

**Keywords:** Blalock-Taussig shunt, hypoplastic left heart syndrome, Norwood procedure.

The first successful use of the Norwood procedure was reported by Norwood et al [1, 2]. Since then, this surgery procedure is most often performed to treat hypoplastic left heart syndrome (HLHS). HLHS is certain types of mitral atresia, which is a lethal condition. The current surgical strategy for HLHS has evolved into a three-stage palliative approach. The first-stage Norwood procedure is performed in the neonatal period. In general, early survival after the Norwood procedure falls in the range between 70-80% [3, 4]. Many institutes have reported increasing survival after using a right ventricular to pulmonary artery (RV-PA) conduit (Sano shunt) [5-7], instead of Blalock-Taussig (BT) shunt. However, according to our experience, we still use only BT shunt with the Norwood procedure.

In this study, we analyzed intermediate results of the treatment of HLHS with the Norwood procedure and BT shunt at the King Chulalongkorn Memorial Hospital since 1996.

**Patients and methods**

**Patients**

Twenty-six patients with HLHS underwent a Norwood procedure at the King Chulalongkorn Memorial Hospital between August 1996 and November 2008. Patients were unselected. The majority had a normal aortic arch (n=21, 80.8%) and the remaining five patients had varying degrees of aortic arch obstruction (Table 1). The median diameter of the ascending aorta was 2.5 mm (range, 2.7 mm). The Norwood procedure was performed at a median age of 11 days (range, 3-75 days). The median weight at operation was 2,850 gm (range, 2,000-4,200 gm).
All operations were performed under deep hypothermic cardiopulmonary bypass (rectal temperature at 18°C) with circulatory arrest. Myocardial protection was provided with a single dose of cold crystalloid cardioplegia (30ml/kg body weight) administered from a side port of the arterial cannula after temporarily occluded arch vessels by tourniquets and by clamping of the descending aorta. The median duration of cardiopulmonary bypass (CBP) and deep hypothermic circulatory arrest (DHCA) time were 96 minutes (range: 51-163 minutes) and 66 minutes (range: 51-97 minutes), respectively. The median period of cardiopulmonary support (the cumulative duration of CBP and DHCA) was 159 minutes (range: 125-216 minutes).

The main pulmonary artery was divided at the level of bifurcation and the distal pulmonary artery was enlarged with a piece of pericardial patch in patients who used autologous tissue for arch reconstruction and with homograft patch in patients whose arch reconstruction was done with homograft. An atrial septectomy was performed during circulatory arrest through the venous cannulation site (right atrial appendage).

The arch was reconstructed using one of the two techniques. The first four patients were operated on by the original technique using only autologous great vessel tissue [8]. All of the remainders were reconstructed by homograft patch since September 1999 (3 aortic homografts, 19 pulmonic homografts). The entire isthmic part of the aorta was removed with adjacent ductal tissue in all patients.

Pulmonary blood flow was established with a right modified Blalock-Taussig shunt (RMBTS). The RMBTS was formed by anastomosing a polytetrafluoroethylene (PTFE) tube graft between the innominate artery and the upper border of the right pulmonary artery. A PTFE 4 mm-graft was used in patients whose weight was more than 3.5 kg, and a 3.5 mm-graft was used in patients whose weight was less than 3.5 kg.

**Postoperative management**

Dopamine at 5-10 μg/kg/min doses was used as standard inotropic agent. Sedation was continued for 12-24 hours postoperatively according to the hemodynamic condition.

**Late management**

Elective cardiac catheterization was performed at four months after the first stage of palliation. The second stage bidirectional Glenn (BDG) procedure was performed five-ten months after the first stage operation. Operation was performed through median sternotomy under cardiopulmonary bypass with beating heart, except in some patients who had interatrial restriction and needed atrial septectomy.

**Statistic analysis**

This was a retrospective study based on the review of hospital records and operative notes. Data were expressed as median (range) and percentage.

**Results**

Outcomes for all patients are summarized in Fig. 1.

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**Table 1.** Number of patients with hypoplastic left heart syndrome (HLHS) without aortic arch obstruction.

<table>
<thead>
<tr>
<th><strong>HLHS without aortic arch obstruction</strong></th>
<th><strong>Number of patients</strong></th>
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<tbody>
<tr>
<td>Various degree of aortic arch obstruction</td>
<td>5 (19.2%)</td>
</tr>
<tr>
<td>Interrupted aortic arch (type B)</td>
<td>1</td>
</tr>
<tr>
<td>Hypoplasia aortic arch</td>
<td>1</td>
</tr>
<tr>
<td>Hypoplasia aortic arch with tricuspid regurgitation</td>
<td>1</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>2</td>
</tr>
</tbody>
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Early mortality and morbidity
Six patients (23.1%) died during the hospitalization for the Norwood procedure. Half of them expired in the operating room. The patients were divided into two groups by the technique of arch reconstruction with or without homograft. Hospital mortality (30-day) in group 1 (arch reconstruction with autologous tissue) was 50.0% and group 2 (arch reconstruction with homograft) was 18.2%, (Table 2).

There were two survivals who developed coarctation of the aorta and had successful treatment with balloon dilatation. One patient developed interatrial restriction and need surgical atrial septectomy.

Late mortality
There were two late deaths (2/26 patients, 7.7%) and 12 months after stage 1 palliation.

Long term survival
Of the 20 survivors (77.9%), there were only five whose pulmonary anatomy and hemodynamic studies were suitable for the BDG procedure. Three of them had the modified Fontan procedure and another two were waiting for the modified Fontan procedure. For progression of cyanosis and unsuitability for the BDG procedure, an additional systemic-to-pulmonary shunt was performed in three survivors. One of them could receive the modified Fontan procedure 12 years later without prior BDG procedure. In summary, four out of twenty survivors (20%) had the modified Fontan procedure.

Discussion
Survival in patients where the arch was reconstructed with homograft was better than when it was performed with only autologous tissue. Most of non-surviving patients had cardiomegaly (demonstrated by chest radiography before operation) and intra-operation because of thick wall in pulmonary arteries. Half of non-surviving patients could not be weaned off cardiopulmonary bypass and expired in the operating room. Recently, many center replaced systemic to pulmonary shunt (BTS) with RV-PA conduit. Many studies demonstrated no difference in early hemodynamic profile between patients

<table>
<thead>
<tr>
<th>Technique</th>
<th>Number of patients</th>
<th>Hospital mortality</th>
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<tbody>
<tr>
<td>Group 1</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Autologous great vessel tissue</td>
<td>4</td>
<td>2 (50.0%)</td>
</tr>
<tr>
<td>Group 2</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Homograft patch</td>
<td>22</td>
<td>4 (18.2%)</td>
</tr>
<tr>
<td>Pulmonic homograft</td>
<td>19</td>
<td></td>
</tr>
<tr>
<td>Aortic homograft</td>
<td>3</td>
<td></td>
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</table>
undergoing a BTS or a RV-PA conduit except for a higher diastolic blood pressure in the RV-PA conduit patent. RV-PA conduit did not improve postoperative survival [9-12]. In our experience, we are still satisfied with the Norwood procedure with RMBTS. There was one survivor who had the modified Fontan procedure without BDG at 12 years old. This patient had an excellent result. Therefore, some of the survivors who were not suitable for the BDG procedure may reach the modified Fontan procedure in the future.

In conclusion, despite limited resource and a small number of patients in this study, the outcome of first stage palliation was in the acceptable range.

The authors have no conflict of interest to report.

References