Fontan completion after Norwood with bidirectional Glenn shunt: their haemodynamics and pulmonary artery configuration

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Abstract

OBJECTIVES: We ranked the haemodynamics and pulmonary artery (PA) configurations of Norwood with bidirectional Glenn shunt (NW-G) patients among the other staged Fontan completion (conventional) patients in this study.

METHODS: Between August 2001 and April 2010, 91 consecutive patients completed staged Fontan operations. Among them, NW-G was performed in 11.

RESULTS: There was no operative or late death except in one case which resulted from a reoperation after Fontan completion. Although the age at bidirectional Glenn (BDG) and Fontan completion was younger in NW-G (4.2 vs. 10.9 months, \( P < 0.01 \), and 2.3 vs. 3.3 years, \( P < 0.01 \)), the interval between BDG and Fontan completion was not significantly different in these groups (23.6 vs. 28.0 months, \( P = 0.71 \)). On Fontan completion, the extracardiac conduit size was significantly different (16.4 vs. 17.7 mm, \( P < 0.01 \)). However, the pulmonary artery size (PA index) and the PA pressure were not significantly different (165.4 vs. 205.1 mm²/m², \( P = 0.23 \), and 13.4 vs. 11.0 mmHg, \( P = 0.08 \)). Particularly in hypoplastic left heart syndrome patients, the PA index was not significantly different between these groups (165.4 vs. 174.9 mm²/m², \( P = 0.59 \)).

CONCLUSIONS: The NW-G patients could have a subsequent Fontan completion with good results. There was no significant difference with respect to Fontan completion.

Keywords: Norwood with bidirectional Glenn shunt • Staged Fontan operation • Haemodynamics • Pulmonary artery configuration

INTRODUCTION

Although the results of the primary Norwood operation in hypoplastic left heart syndrome (HLHS) have markedly improved, postoperative mortality remains \(~10–20\% [1, 2]\).

An alternative less invasive stage I palliation, bilateral pulmonary artery banding (bPAB), was introduced, especially in high-risk HLHS patients. The advantage of bPAB is to avoid extracorporeal circulation in the early neonatal period. bPAB could also avoid balancing delicate changes in resistance of the systemic and PA just after primary Norwood palliation. It was preferable to reduce the right ventricular preload by controlling the pulmonary flow. However, maintenance of a wide patent pulmonary artery-ductus-descending aorta-trunk (PDDT) as the systemic perfusion source and sufficient inter-atrial communication was essential between the bPAB and subsequent Norwood operation; thus, interstage management may be complicated.

We introduced bPAB in 2001 in patients with high-risk HLHS and its variants. From 2003, we performed bPAB in all patients who needed the Norwood-type operation. After bPAB, a Norwood with bidirectional Glenn shunt (NW-G) was performed in 14 patients who could wait >4 months, which was generally the earliest possible time with the BDG physiology [3–5], and weighed >4 kg. Among them, 11 patients (73%) completed the Fontan operation. On the other hand, for the patients who could not meet the criteria, we performed a Norwood with the systemic–pulmonary shunt. This strategy had some concerns relating to branch PAs. It is possible that the central PA could deform at the banding site, and reduced pulmonary flow by bPAB with the subsequent BDG physiology could not grow the PA tree.

The purpose of this study was to rank the haemodynamics and PA configuration of the NW-G group among groups of other conventional procedures in staged Fontan completions, which essentially are of a heterogeneous nature.

PATIENTS AND METHODS

Between August 2001 and April 2010, 91 consecutive patients completed staged Fontan operations at our hospital (Table 1). Among them, NW-G with initial palliation by bPAB was performed in 11. The other patients (\( n = 80 \)) were classified as the conventional group.
Table 1: Patient profiles (n = 91)

<table>
<thead>
<tr>
<th>NW-G</th>
<th>n = 11</th>
</tr>
</thead>
<tbody>
<tr>
<td>HLHS</td>
<td>8</td>
</tr>
<tr>
<td>DILV with IAA</td>
<td>2</td>
</tr>
<tr>
<td>CCH with CoA</td>
<td>1</td>
</tr>
<tr>
<td>Conventional</td>
<td>n = 80</td>
</tr>
<tr>
<td>HLHS</td>
<td>11</td>
</tr>
<tr>
<td>DIRV</td>
<td>9</td>
</tr>
<tr>
<td>DILV</td>
<td>1</td>
</tr>
<tr>
<td>TA</td>
<td>7</td>
</tr>
<tr>
<td>Ebstein</td>
<td>2</td>
</tr>
<tr>
<td>PAIVS, severe PS</td>
<td>11</td>
</tr>
<tr>
<td>CCH</td>
<td>3</td>
</tr>
<tr>
<td>CAVC with unbalanced ventricles</td>
<td>10</td>
</tr>
<tr>
<td>Multiple VSDs</td>
<td>8</td>
</tr>
<tr>
<td>DORV + unbalanced ventricles</td>
<td>11</td>
</tr>
<tr>
<td>DORV + remote VSD</td>
<td>1</td>
</tr>
<tr>
<td>DORV + straddling AVV</td>
<td>3</td>
</tr>
<tr>
<td>ccTGA + unbalanced ventricles</td>
<td>2</td>
</tr>
<tr>
<td>ccTGA + straddling AVV</td>
<td>1</td>
</tr>
</tbody>
</table>

NW-G: Norwood with bidirectional Glenn shunt; HLHS: hypoplastic left heart syndrome; DILV: double inlet left ventricle; IAA: interruption of aortic arch; CCH: criss-cross heart; CoA: coarctation of aorta; DIRV: double inlet right ventricle; TA: tricuspid atresia; PAIVS: pulmonary atresia with the intact ventricular septum; CAVC: common atrioventricular canal; VSD: ventricular septal defect; DORV: double outlet right ventricle; AVV: atrioventricular valve; ccTGA: congenitally corrected transposition of the great arteries.

Bilateral pulmonary artery banding

bPAB as an initial palliation was performed in all NW-G patients. This strategy required maintaining the PDDT and inter-atrial communication as a source of systemic perfusion until the Norwood operation. We routinely used continuous intravenous drip infusion of prostaglandin E1 and percutaneous balloon atrial septotomy. Only two patients underwent stent implantation in PDDT in this series. The mean age at bPAB was 7.5 days in this series. All but one patient was treated with nitrogen gas inhalation to control pulmonary overcirculation before bPAB. The banding tape material was a 2-mm-wide strip made of an expanded polytetrafluoroethylene graft. The tightness of the bilateral bands was adjusted to a PO2 of 35–40 mmHg with room air and normocapnia. In our series, each banding tape length was predetermined to be ~0.6–0.7 times as long as the measured circumference of the branch PA. As a result, the mean length of banding tape was 9.1 ± 1.5 mm for the right PA and 8.6 ± 1.3 mm for the left PA. Direct epicardial echocardiography was also used to evaluate the branch PA pressure beyond the banding site. Peak velocity was 3–3.5 m/s at the banding site.

Norwood with bidirectional Glenn shunt

After the initial palliation by bPAB, NW-G as a stage 2 palliation was performed in patients who met the following criteria: more than 4 months of age, larger than 4 kg, because BDG can be performed more safely after 3 months of age [3–5]. NW-G consisted of reconstruction of the neoaoortic arch and PA. To avoid circulatory arrest during arch repair, we routinely used the descending aortic perfusion in addition to the brachiocephalic artery perfusion.

The bilateral branch PAs were debanded, and gently dilated with a Hegar dilator through the transected pulmonary stump. Only this manoeuvre was performed in all but two patients. Left PA disruption occurred in one patient and required left PA angioplasty. Intraoperative direct balloon catheter angioplasty was performed in one patient for prophylaxis against pulmonary stenosis.

Staged Fontan completion

In general, we performed the staged Fontan operation using an extracardiac conduit without fenestration in single-ventricle lesion patients [6]. A systemic–PA shunt operation or banding of the main pulmonary trunk was performed in the neonatal or early infancy period if needed. BDG was usually performed in infants aged 6 months or older in our hospital, if there was no possibility of biventricular repair. In borderline patients with unbalanced ventricles, a longer and more careful observation period was needed before BDG. Close observation is needed during the BDG and Fontan operation, because it is widely believed that too long a waiting period can result in the narrowing of the PA and the formation of a pulmonary arteriovenous fistula [7, 8]. As for the timing of the Fontan completion, physical size was an important criterion, as well as haemodynamics. In patients weighing over 10 kg, a 16-mm or larger extracardiac conduit could be used to avoid re-operation associated with the somatic growth. Post-Fontan catheterization data were obtained from investigation at 1-year after a Fontan completion. We confirmed that there was no significant pressure gradient in PAs.

Statistical analysis

Preoperative and postoperative data were analysed between the groups. Data were expressed as the mean ± SD. The significance of differences between the two groups was determined by the two-tailed Mann–Whitney U-test. A P-value <0.05 was considered significant. Statistical analysis was performed with the GraphPad Prism 5.0 Software package.

RESULTS

Bilateral pulmonary artery banding and subsequent Norwood with bidirectional Glenn shunt

This study was focused on the patients who completed the staged Fontan operation between August 2001 and April 2010. During this period, bPAB for the Fontan operation was performed in 15 patients (HLHS in 12, single ventricle with interrupted aortic arch in two and criss-cross heart with coarctation of the aorta in one). After bPAB, one patient died of uncontrollable severe tricuspid insufficiency in HLHS. The remaining 14 patients underwent NW-G. Among them, 11 completed the Fontan operation and the other three were waiting for the Fontan operation.

Bidirectional Glenn shunt

There were no early or late deaths after BDG in either groups. The NW-G operation was performed at 4.2 ± 0.6 months of age. On
the other hand, BDG was performed at 20.9 ± 21.3 months of age in the conventional group. In 23 patients, biventricular repair had to be abandoned because of unbalanced ventricles, and they had a longer observation period before BDG. Excluding those cases, BDG was performed at 10.9 ± 5.2 months of age (Table 2).

### Fontan completion

There were no early or late deaths after Fontan completion in either groups, except for one patient, which resulted from re-operation of the common atroventricular valve 4 years after a Fontan completion. The interval between BDG and Fontan was not significantly different between the groups (23.6 ± 5.1 months in the NW-G vs. 25.2 ± 14.9 months in the non-NW-G, P = 0.71). Consequently, age at Fontan was significantly younger (2.3 ± 0.4 vs. 3.8 ± 1.8 years, P < 0.0001), body weight was significantly lighter (10.4 ± 0.9 vs. 13.4 ± 3.1 kg, P < 0.0001) and a significantly smaller conduit (16.4 ± 0.8 vs. 17.7 ± 0.8 mm, P < 0.0001) was implanted in the NW-G group (Table 2).

Excluding 23 patients in whom biventricular repair had to be abandoned owing to unbalanced ventricles in the conventional group, the results showed the same tendency. The interval between BDG and Fontan was not significantly different between the groups (23.6 ± 5.1 vs. 28.0 ± 14.5 months, P = 0.35). As a consequence, age at Fontan was significantly younger (2.3 ± 0.4 vs. 3.3 ± 1.2 years, P < 0.01), body weight was significantly lighter (10.4 ± 0.9 vs. 12.5 ± 1.5 kg, P < 0.01) and a significantly smaller conduit (16.4 ± 0.8 vs. 17.7 ± 0.8 mm, P < 0.01) was used in the NW-G group (Table 2).

### Pulmonary artery configuration and pulmonary artery pressure

The PA index [9] was not significantly different between the groups (154.0 ± 35.1 vs. 209.4 ± 80.0 mm²/m², P = 0.12 in pre-Fontan, and 165.4 ± 44.0 vs. 205.1 ± 73.3 mm²/m², P = 0.23 in post-Fontan).

The PA pressure in pre-Fontan patients was not significantly different between the groups (9.2 ± 1.7 vs. 8.9 ± 1.7 mmHg, P = 0.54). Although the PA pressure in post-Fontan patients was elevated in both the groups, there was no significant difference between the groups (13.4 ± 4.0 vs. 11.0 ± 2.5 mmHg, P = 0.08).

### Cardiac function

The ejection fraction as an index of systemic ventricular performance was not significantly different between the groups (57.5 ± 3.2 vs. 59.5 ± 9.7% in pre-Fontan, and 55.4 ± 7.7 vs. 58.9 ± 10.1% in post-Fontan).

### Hypoplastic left heart syndrome

As for the timing of BDG, there was no significant difference between the groups (4.0 ± 0.4 vs. 6.4 ± 4.1 months, P = 0.08). The interval between BDG and Fontan was significantly shorter in NW-G (25.0 ± 4.4 vs. 37.7 ± 12.5 months, P = 0.008). As a result, the Fontan operation was performed significantly earlier (2.4 ± 0.4 vs. 3.7 ± 1.0 years, P < 0.01), and the body weight at Fontan was significantly lower (10.7 ± 0.8 vs. 12.4 ± 1.1 kg, P < 0.01). However, the implanted conduit size was not significantly different between the groups (16.5 ± 0.9 vs. 17.3 ± 1.0 mm, P = 0.12). The PA index was not significantly different between the groups (158.8 ± 31.0 vs. 167.7 ± 73.3 mm²/m², P = 0.45 in pre-Fontan, and 165.4 ± 44.0 vs. 174.9 ± 41.8 mm²/m², P = 0.59 in post-Fontan, Table 3).

### DISCUSSION

There have been no early or late deaths with the combination of bPAB and subsequent NW-G, thus far, except in one that

<table>
<thead>
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<th>Table 2: Results</th>
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<tr>
<td>NW-G (n = 11)</td>
</tr>
<tr>
<td>Age at BDG (months)</td>
</tr>
<tr>
<td>Age at Fontan (years)</td>
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<tr>
<td>BW at Fontan (kg)</td>
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<td>Interval between BDG and Fontan (months)</td>
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<td>Conduit (mm)</td>
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<td>PA index, pre-Fontan (mm²/m²)</td>
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<td>PA index, post-Fontan (mm²/m²)</td>
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<td>PAp, pre-Fontan (mmHg)</td>
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<td>PAp, post-Fontan (mmHg)</td>
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<tr>
<td>EF, pre-Fontan (%)</td>
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<td>EF, post-Fontan (%)</td>
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<td>SpO₂, pre-Fontan (%)</td>
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*Excluding 23 BDG cases who had to abandon biventricular repair because of unbalanced ventricles. NW-G: Norwood with bidirectional Glenn shunt; BDG: bidirectional Glenn shunt; BW: body weight; PA index: pulmonary artery index; PAp: pulmonary artery pressure; EF: ejection fraction; SpO₂: percutaneous oxygen saturation.
resulted from an uncontrollable severe systemic atrioventricular valve insufficiency in HLHS. The primary Norwood operation has 10% mortality at best [1, 2]. An invasive surgical procedure using extracorporeal circulation in the neonatal period, and post-operative disturbance of delicate systemic-pulmonary flow balance, which can worsen right ventricular function, are responsible for the outcome. bPAB and subsequent NW-G could prevent both the use of extracorporeal circulation in the neonatal period and systemic-pulmonary flow imbalance after the Norwood operation.

Instead, maintaining the PDDT and inter-atrial communication is a critical issue in the interstage management. Although stent placement in the PDDT and the atrial septum is emerging as an alternative strategy [10], we prefer the continuous use of prostaglandin E1, because stent placement in PDDT could complicate arch reconstruction with autologous tissue in the subsequent Norwood operation.

The NW-G operation was performed at ~3 or 4 months of age. On the other hand, many BDG operations are usually performed at 4 months or older at the earliest. In our series, biven-tricular repair had to be abandoned in 23 patients because of borderline unbalanced ventricles in the conventional group. With the exception of those cases, BDG was performed at 10.9 ± 5.2 months of age. There was a significant difference in the timing of BDG between the groups. The interval between BDG and Fontan completion was not different between the groups, and therefore the mean age, mean body weight and mean conduit size were significantly smaller at Fontan completion in the NW-G group.

**Fontan completion**

As to the timing of Fontan completion, there are different opinions [11]. The Fontan completion rate through initial palliation by bPAB and subsequent NW-G was excellent. From the standpoint of the survival rate and the Fontan completion rate, our strategy contributed greatly to the improved surgical outcome.

In HLHS, the NW-G group had Fontan completion significantly earlier than the conventional group. There was a bias in patient selection, as NW-G was performed only in patients who could meet the criteria. As a result, they could undergo a straightforward Fontan completion.

**Pulmonary configuration**

The PA index and the PA pressure were not significantly different between the groups. This suggested that bPAB and subsequent NW-G did not affect PA growth. However, our particular concern was that the accumulation of operations for low pulmonary flow might be impedimental to PA growth. bPAB aimed to reduce PA flow. Also, subsequent BDG operation never resulted in the high pulmonary flow [7].

Nakano et al. [12] reported that bPAB did not cause later PA stenosis in the HLHS patients. In our HLHS patients, there was no difference in haemodynamics and PA configuration between the groups. This suggests that bPAB did not affect PA growth. Rather, the Norwood-type operation could impair PA growth, because compression within the aortopulmonary space after reconstruction of the aorta could result in PA narrowing [13].

**Study limitation**

This study was retrospective, from a single-centre, and non-randomized. Because our patient classification was not based on risk stratification, it was possible that there was a bias especially in HLHS patient selection between the groups.

**CONCLUSION**

The NW-G patients following bPAB could have a complete subsequent Fontan operation with good results. There was no significant difference in PA configuration and haemodynamics between the NW-G and conventional groups. Especially in the HLHS patients, there was no significant difference in the PA index and the PA pressure. bPAB and the subsequent NW-G operation did not affect PA growth. Further observation of the PA configuration should be conducted.
Conflict of interest: none declared.

REFERENCES


