Institutional report - Congenital

Stage I bilateral pulmonary artery banding maintains systemic flow by prostaglandin E1 infusion or a main pulmonary artery to the descending aorta shunt for hypoplastic left heart syndrome

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1. Introduction

In most institutions, the Norwood procedure for stage I palliation of hypoplastic left heart syndrome (HLHS) is the procedure of choice. Though an improved outcome of this staged approach has been reported [1], the mortality rate after stage I Norwood is still high [1,2]. The Norwood operation is an invasive procedure, usually required during the neonatal period. The balance between the systemic and pulmonary resistance in this period may easily induce hemodynamic instability after the Norwood procedure. In our institution we perform a less invasive operation for hemodynamic instability after the Norwood procedure. In stage II palliation, the arterial line of the cardiopulmonary bypass circuit was divided in two into a Y shape; one branch was used for cerebral perfusion through the innominate artery and the other for lower body perfusion through a cannula inserted into the descending thoracic aorta.

2. Subjects and methods

Four consecutive patients with ‘classic’ HLHS underwent the bilateral PA banding strategy from January 2002 to April 2003 at the Department of Thoracic and Cardiovascular Surgery, Mie University School of Medicine. We retrospectively reviewed charts of these 4 HLHS patients, obtaining data regarding: cardiac anatomy, age and weight at the staged operation, preoperative treatment, diameter of bilateral PA banding, oxygen saturation (Sat O₂), systemic flow maintenance method, interstage status, right pulmonary artery mean pressure (RPAPm), PA index (PAI) defined right PA area (mm²) + left PA area (mm²)/body surface area (m²) [7], additional procedure of stage II palliation, demographics, operation time, cardiopulmonary bypass (CPB) time, aorta (Ao) clamp time, possibility of sternal closure, mechanical ventilation, dopamine duration, chest drainage, pulmonary artery mean pressure (PAPm), and reports of surgeries, cardiac catheterizations, and echocardiography. In stage II palliation, the arterial line of the cardiopulmonary bypass circuit was divided in two into a Y shape; one branch was used for cerebral perfusion through the innominate artery and the other for lower body perfusion through a cannula inserted into the descending thoracic aorta.

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Abstract

Since 2002, we have performed bilateral pulmonary artery banding for stage I palliation and maintained systemic flow by prostaglandin E1 infusion or a main pulmonary artery to the descending aorta shunt, and here report our experience. Three of the 4 patients were diagnosed with aortic atresia/mitral atresia and 1 with aortic stenosis/mitral stenosis. Balloon atrial septostomy was performed in 2 before stage I. Bilateral pulmonary artery banding (right circumference: 10 or 14, left circumference: 10.5 to 14 mm) was performed from 7 to 19 days after birth. Systemic flow was maintained by prostaglandin E1 infusion in 2 patients and a Van Praagh procedure was performed in the other 2. Balloon atrial septostomy was required in 2 patients, and an atrial septal defect enlargement was in one during the interstage before stage II palliation, which was performed at ages 3 to 9 months. Bidirectional cavopulmonary shunt with aortic arch and coronary flow reconstruction was also performed. For patients younger than 4 months, we do not require pulmonary arterioplasty in stage II. All patients are alive and well and waiting for Fontan completion. Excellent early results were obtained for this surgical strategy that avoids the stage I Norwood palliation.

Keywords: Hypoplastic left heart syndrome; Bilateral pulmonary artery banding; Stage I palliation; Less invasive surgery; Staged Fontan; Norwood procedure
This retrospective review was approved by our institution in May 2004.

3. Results

Four neonatal HLHS patients were diagnosed: 3 aortic atresia/mitral atresia and 1 aortic stenosis/mitral stenosis. Ages at stage I palliation were 7 to 19 days (weight 2.6 to 3.8 kg). Balloon atrial septostomy (BAS) was performed in 2 patients at 4 and 6 days before bilateral PA banding; no sudden increase in pulmonary blood flow was experienced in either patients after BAS. Mechanical ventilation was used in 3 preoperatively. Nitrogen treatment ($N_2$) was used for all patients to control oxygen saturation to approximately 80%. Bilateral PA banding was performed to create a circumference on the right of 10 or 14 mm and on the left 10.5 to 14 mm by our hand-made banding tape in an ePTFE sheet of 0.4 mm thickness was trimmed to 2 mm width. We decided the circumference of the bilateral PA banding mainly from the parameter of Sat $O_2$. In the last 2 patients, we performed the bilateral PA banding of a circumference of 11 mm and an additional closing of 0.5 mm to have Sat $O_2$ between 70 and 80%. Maintenance of systemic flow was performed with a main pulmonary artery to the descending aorta shunt via a left thoracotomy (Van Praagh graft; ePTFE 6 mm) in patients 1 and 2, and by PGE1 administration (5 ng/kg/min) in patients 3 and 4. In the PGE1 patients, early extubation was possible (patient 3: 2 days after the stage I palliation, patient 4: the operative day); by contrast, the patients receiving a Van Praagh procedure required longer mechanical ventilation (patient 1: 49 days, patient 2: not extubated until stage II palliation). In both the PGE1 patients, there was no problem with venous access in the period between stages I and II palliation. Bilateral PA banding was performed through a left thoracotomy in the patients using the Van Praagh procedure, or through a median sternotomy in patients using PGE1 without CPB (Table 1). The sternum was closed in all cases.

The PGE1 patients were managed in the neonatal intensive care unit until stage II palliation was performed at 3 or 4 months of age. Stage II palliation was performed in the Van Praagh procedure patients at 7 or 9 months of age when their weight was 3.5 to 4.7 kg. In the interstage between stage I palliation and stage II palliation, the restrictive atrial septal defect (ASD) progressed in all patients. BAS was performed in 2, ASD enlargement was performed in one, and one had a restrictive ASD and an enlarged ASD at stage II palliation, concomitantly. The ductus arteriosus was kept patent in all patients, including those with previous ductal shock. Two patients had moderate and one mild tricuspid regurgitation (TR). The highest preoperative RPAPm found was 35 mmHg in the patient with restrictive ASD at stage II palliation; the other patients showed an RPAPm of 12 to 16 mmHg. Left pulmonary artery mean pressure (LPAPm) was assessed by pressure gradient on transthoracic echocardiography. The LPAPm ranged from 35 to 55 mmHg. Stage II palliation procedures entailed BCPS and aortic arch and coronary flow reconstruction. Additionally, we performed an additional PA plasty in 2 patients and tricuspid annuloplasty in 2 patients (Table 2). The sternum was closed in all cases.

The latest postoperative PAP was 12, 13, 12, and 14 mmHg and the PAI was 110, 111, 120, 135 mmHg/m² in patients 1 to 4, respectively. Presently, all patients who underwent the Norwood procedure and BCPS are alive and well at 28, 27, 23, and 24 months of age, respectively, and waiting for Fontan completion (Fig. 1).

### Table 1
Patient characteristics – stage I palliation

<table>
<thead>
<tr>
<th>Patient</th>
<th>Diagnosis</th>
<th>Age (d)</th>
<th>Weight (kg)</th>
<th>Ductal shock</th>
<th>BAS-</th>
<th>Mechanical ventilation</th>
<th>$N_2$</th>
<th>Circumference of BPAB (mm)</th>
<th>Pre SAT $O_2$(%)</th>
<th>Post SAT $O_2$(%)</th>
<th>Maintenance of systemic flow</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>AA/MA mild TR</td>
<td>19</td>
<td>2.6</td>
<td>n</td>
<td>y</td>
<td>y</td>
<td>y</td>
<td>14/14</td>
<td>88</td>
<td>82</td>
<td>VP</td>
</tr>
<tr>
<td>2</td>
<td>AA/MA mild TR bil.SVC</td>
<td>15</td>
<td>3.0</td>
<td>n</td>
<td>n</td>
<td>y</td>
<td>y</td>
<td>14/14</td>
<td>85</td>
<td>81</td>
<td>VP</td>
</tr>
<tr>
<td>3</td>
<td>AA/MA</td>
<td>16</td>
<td>3.8</td>
<td>y</td>
<td>y</td>
<td>y</td>
<td>y</td>
<td>10/11.0</td>
<td>88</td>
<td>78</td>
<td>PGE1</td>
</tr>
<tr>
<td>4</td>
<td>AS/MS mild TR</td>
<td>7</td>
<td>3.5</td>
<td>n</td>
<td>n</td>
<td>y</td>
<td>y</td>
<td>10/10.5</td>
<td>93</td>
<td>73</td>
<td>PGE1</td>
</tr>
</tbody>
</table>

* Treatment preoperatively. † With $N_2$. ‡ Room air. BAS = balloon atrial septostomy; $N_2$ = nitrogen; BPAB = bilateral pulmonary artery banding; Sat $O_2$ = oxygen saturation; Pre = preoperative; Post = postoperative; AA = aortic atresia; MA = mitral atresia; TR = tricuspid regurgitation; n = no; y = yes; VP = VanPrag; bil.SVC = bilateral superior vena cava; PGE1 = prostaglandin E1; MS = mitral stenosis.

### Table 2
Patient characteristics – stage II palliation

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (m)</th>
<th>Weight (kg)</th>
<th>Restrictive ASD*</th>
<th>Ductus patency*</th>
<th>TR*</th>
<th>RPAPm (mmHg)</th>
<th>PA index (mm²/m²)</th>
<th>Additional procedure*</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>9</td>
<td>4.5</td>
<td>ASD enlargement</td>
<td>y</td>
<td>Moderate</td>
<td>12</td>
<td>746</td>
<td>PA plasty TAP</td>
</tr>
<tr>
<td>2</td>
<td>7</td>
<td>3.5</td>
<td>y</td>
<td>y</td>
<td>Mild</td>
<td>35</td>
<td>541</td>
<td>PA plasty</td>
</tr>
<tr>
<td>3</td>
<td>4</td>
<td>4.7</td>
<td>BAS</td>
<td>y</td>
<td>n</td>
<td>15</td>
<td>151</td>
<td>n</td>
</tr>
<tr>
<td>4</td>
<td>3</td>
<td>4.5</td>
<td>BAS</td>
<td>y</td>
<td>Moderate</td>
<td>16</td>
<td>168</td>
<td>TAP</td>
</tr>
</tbody>
</table>

* Preoperative status. † At stage II palliation. TR = tricuspid regurgitation; RPAPm= right pulmonary artery mean pressure; PA = pulmonary artery; ASD = atrial septal defect; n = no; y = yes; TAP = tricuspid annuloplasty; BAS = balloon atrial septostomy.
Stage II palliation, that is from unexpected death in a home environment. The effective reduction of the interstage mortality before hospitalization, the merits of maintaining hospitalization is the cost of keeping patients on PGE1 infusion and in chronic ICU hospitalization; this is because of the high interstage mortality due to hemodynamic problems in HLHS patients. Although some two million yen per month of age before ductus closure to stabilize patients with acidosis [3] or with an obstructed anomalous pulmonary to systemic venous connection between the left atrium and the innominate vein [4]. Akintuerk [5] reported a combination of bilateral PA banding and stenting for ductus.

Our ideal treatment for HLHS is; before bilateral PA banding, BAS for restrictive ASD performed using 

An important issue is restrictive ASD [13], which in our experience, was progressive in all patients until stage II palliation. ASD enlargement was performed in 2 patients before stage I palliation and in 3 patients during the interstage before stage II palliation. To correctly perform bilateral PA banding, the ASD must be open and so reduce postoperative mortality. High pulmonary vascular resistance at stage II palliation from the progression of a severely restrictive ASD would prevent a successful BCPS completion. Advanced catheter techniques including BAS or a stent would be required for certain restrictive ASDs.

Interestingly, the ductus of all patients after bilateral PA banding were patent. This finding suggests that the increased systemic flow through the ductus arteriosus effects patency. None of our patients had coarctation that would have prevented adequate back flow to the aortic arch, head vessels, and coronary arteries. Although we think the ductal opening would be maintained by an increased flow through the ductus after banding with PGE1, it is unclear whether it might stay patent without PGE1. Physiologic and histologic study of ductus arteriosus should make the mechanisms involved in ductus arteriosus patency clearer. We recommend maintenance of systemic flow by PGE1 administration, because ductal stenting [5] can be accompanied by complications of injury and iatrogenic narrowing of the ductus arteriosus. Ductus arteriosus tissue is weak and easily injured; PGE1 administration naturally formed a patent ductus arteriosus. The progression of ductal narrowing, despite PGE1 treatment would necessitate a Van Praagh procedure via a left thoracotomy. Our experience is that Van Praagh as a route to systemic circulation is a useful technique.

In the two patients having BCPS at 7 and 9 months, the PA tissue at the banding site had become fragile. Therefore, PA plasty was required in both patients. In the PGE1 patients with early stage II palliations performed at 3 and 4 months of age, the release of adhesions was easier, and aortic arch reconstruction was performed with the patient’s own aortic and pulmonary vascular tissue, so that PA plasty was not required. Further, it would appear that performing a combined stage II operation at a younger age reduces the volume-load on the ventricle and improves oxygen saturation. In HLHS patients, TR [14] is the most common issue; if neonatal progressive TR occurred, we would perform bilateral PA banding and tricuspid annuloplasty concomitantly without aortic arch reconstruction. A hypoplastic ascending aorta correlated with mortality after a Norwood procedure [15], although reduced coronary flow before stage II palliation was not found in any patients.

The limitation of this study is the small number of patients, and the methods of systemic flow maintenance and banding circumference are different between each set of 2 patients. Further study of larger numbers will be required to gauge the adequacy of this treatment for stage I palliation and interstage management.

In conclusion, the results of our bilateral PA banding strategy for HLHS were excellent. Bilateral PA banding for stage I palliation with systemic flow maintained by PGE1 infusion without CPB is a less invasive protocol that may reduce early stage I palliation mortality. The Norwood procedure and BCPS without circulatory arrest reduced the...
volume overload and promoted hemodynamic stability. Thus postoperative management at stage I palliation and stage II palliation was easier. The advantages detailed here will promote a lower mortality rate of staged Fontan for HLHS. Therefore, our bilateral PA banding strategy may offer a staged surgical option for HLHS patients.

**References**


**Appendix A. ICVTS on-line discussion**

**Author:** Murat Basaran (GATA Military Training Hospital, Turkey)

**eComment:** Although the morbidity and mortality rates associated with hypoplastic left heart syndrome (HLHS) decreased significantly, this anomaly is still a challenging issue for pediatric cardiac surgeons. In the paper, Takabayashi reported his experience with a technique described by Van Praagh for the correction of interrupted aortic arch. However, with increasing experience, we learned that HLHS also has negative effects on the cerebral system by altering blood flow patterns. We know that the retrograde cerebral circulation may have implications for later development of neurological sequelae and even structural brain abnormalities. The stage I palliation of HLHS with classical Norwood procedure, not only corrects the hypoplastic ascending aorta, but also establishes an antegrade cerebral circulation which is an important factor for brain development. In the described technique, the retrograde cerebral circulation continued until the stage II palliation; therefore, I think that the neurodevelopmental status of the patients at the time of subsequent procedures should also be mentioned in the article by the authors. Lastly, congratulations to the authors in dealing with such a complex cardiac anomaly.

**Author:** Shin Takabayashi (Mie University School of Medicine, Japan)

**eResponse:** Thank you for your eComment. In PGE1 patients, there was no pressure gradient between aortic arch and descending aorta until the stage II palliation and obvious neurodevelopmental impairment was not found. However, 10 mmHg pressure gradient developed at the stage II palliation in one VP patient due to shrinking of ductal tissue. We think that PGE1 management is superior to VP management in cerebral perfusion.