Impact of 3-mm Blalock–Taussig shunt in neonates and infants with a functionally single ventricle

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Abstract

Functionally single ventricle (f-SV) is susceptible to volume overload. Atrioventricular valve regurgitation (AVVR) tends to develop and ventricular function deteriorates due to excessive pulmonary blood flow following modified Blalock–Taussig shunt (mBTS). On the other hand, a small caliber graft has risks of early obstruction and poor growth of pulmonary vascular beds. We assessed the effect of mBTS with a 3-mm graft to circumvent volume overload in f-SV on achievement of the right heart bypass. Eleven neonates and infants with f-SV at the median age of 24 days underwent mBTS using a 3-mm graft between August 2004 and June 2007. There were no early deaths, but there was one late death. All survivors achieved bidirectional cavopulmonary shunt and can thus lead to ventricular dys-function [1–3]. On the other hand, BTS with a small graft may develop shunt occlusion in an early stage and lead to poor growth of pulmonary vascular beds. As a consequence, patients would fail to be good candidates for right heart bypass operation [4, 5]. Recent improvements in the Norwood operation, however, have demonstrated that a 3-mm graft is useful in maintaining ventricular function and thus achieving a right heart bypass operation in patients with hypoplastic left heart syndrome [6, 7]. There was also a report showing no significant differences in size of shunt in pulmonary artery (PA) growth [8]. These facts encouraged us to consider a 3-mm graft for non-open heart palliation in patients with a functionally single ventricle. No studies have focused on the usefulness and effectiveness of BTS using a 3.0-mm graft, although several reports have used a 3.5-mm or larger graft. Therefore, in this study, we focused on a group with a 3.0-mm graft. The purpose of this study was to demonstrate the effectiveness of a 3-mm BTS on ventricular function and pulmonary vascular growth, and to assess the achievement of a right heart bypass operation.

1. Introduction

Functionally single ventricle is susceptible to volume overload. Excessive pulmonary flow through a large Blalock–Taussig shunt (BTS) may worsen atrioventricular valve regurgitation (AVVR) and can thus lead to ventricular dysfunction [1–3]. On the other hand, BTS with a small graft may develop shunt occlusion in an early stage and lead to poor growth of pulmonary vascular beds. As a consequence, patients would fail to be good candidates for right heart bypass operation [4, 5]. Recent improvements in the Norwood operation, however, have demonstrated that a 3-mm graft is useful in maintaining ventricular function and thus achieving a right heart bypass operation in patients with hypoplastic left heart syndrome [6, 7]. There was also a report showing no significant differences in size of shunt in pulmonary artery (PA) growth [8]. These facts encouraged us to consider a 3-mm graft for non-open heart palliation in patients with a functionally single ventricle. No studies have focused on the usefulness and effectiveness of BTS using a 3.0-mm graft, although several reports have used a 3.5-mm or larger graft. Therefore, in this study, we focused on a group with a 3.0-mm graft. The purpose of this study was to demonstrate the effectiveness of a 3-mm BTS on
use was determined intra-operatively on the basis of anas-
tomotic PA size first and proximal systemic artery size
second. When the size of PA at its anastomotic site was
>4 mm, we used a 3.0-mm graft. When it was <4 mm,
we used a 3.5-mm graft.

A 3-mm-ePTFE graft was interposed between the subclav-
ian artery and the PA in nine patients. In two patients it
was interposed between the carotid artery and the PA. The
runoff of the PA was mainly dependent on the extent
of the incision on the subclavian artery. When it was likely
to have good runoff because the pulmonary artery was well
grown, we incised the subclavian artery rather distally. In
contrast, when the PA was small, we extended the incision
partly onto the brachiocephalic artery. We made sure that
the graft was anastomosed to the pulmonary artery as
proximally as possible. Concomitantly, the pulmonary trunk
was banded in one patient and ligated in one patient when
major antegrade flow to the lungs was present. The duct
was ligated in one patient (left-side shunt) and banded in
two patients (median sternotomy in 1, left-side shunt in 1).
The major aortopulmonary collateral arteries were
ligated in one patient.

### 2.3. Postoperative management protocol

Patients were deeply sedated and paralyzed for the first
24 h. The median duration of postoperative mechanical
ventilatory support was three days. Oxygen was used in the
early postoperative period depending on the presumed
changes in pulmonary resistance after surgery. Anticoag-
ulation therapy was started after the BTS procedure. Heparin
infusion commenced immediately after bleeding was iden-
tified as minimal and continued for five days. Aspirin was
initiated when starting oral feeding.

### 2.4. Statistical analysis

The continuous data in this study are expressed as mean
values ± S.D. or as median values. Analyses were performed
using SPSS software (Version 11.0J; SPSS Inc., Chicago, IL,
USA). The probability of BCPS achievement was calculated
using the Kaplan–Meier method.

This study was approved by the Ethics Committee of the
Kanagawa Children’s Medical Center.

### 3. Results

#### 3.1. Fate of the duct and postoperative course

To avoid excessive pulmonary blood flow after surgery,
we administered prostaglandin preoperatively so that it
would spontaneously close soon after creating the BTS.
Therefore, we changed PGE, to prostaglandin CD at one
week before surgery. Echocardiogram showed that the size
of the duct started to decrease between 6 and 24 h after
discontinuing prostaglandin CD. Meanwhile, the blood flow
through the graft started to increase during the same
period. Thereafter, the duct was completely closed from 5
to 12 days after surgery in most patients. At discharge,
median percutaneous oxygen saturation was 80% (70–90%).
Echocardiogram at discharge revealed that AVVR was trivi-
al, or none, in all patients. There were no instances of
stenosis in the central pulmonary artery.

#### 3.2. Graft thrombosis

There was a prolonged hospital stay in two patients; both
of whom incurred acute graft thrombosis at 50 and 81 days
after surgery, respectively; which was successfully treated
by emergency catheter thrombolysis therapy. Both had a
tendency for graft thrombosis.

One had pulmonary atresia with intact ventricular septum
associated with Protein-C deficiency characterized by
hypercoagulability. A diagnosis of Protein-C deficiency was
yet to be made when the graft was occluded. Another
patient had a univentricular heart associated with both
VATER association and bronchomalacia. High end-expiratory
pressure was needed to avoid airway obstruction in the
expiratory phase. The patient recovered from the event,
but died of sepsis following respiratory infection at 96 days
after modified BTS.
3.3. Achievement of right heart bypass

Cardiac catheterization before BCPS is shown in Table 2. Atrioventricular valve regurgitation remained at a level of mild or less and the ventricular function was kept within the normal range with ejection fraction at 63 ± 6%, ventricular end-diastolic volume of 186 ± 71% of normal and ventricular end-diastolic pressure of 7 ± 3 mmHg. Pulmonary angiography showed excellent growth of the PA without any stenosis (Fig. 1).

All survivors accomplished BCPS at 2.0–7.8 months (median, 4.0 months) after 3 mm BTS (Fig. 2) and one patient after another shunt, as mentioned above. The age at BCPS ranged from 3.0 to 10.0 months (median, 5.5 months). Concomitant procedures included TAPVD repair in two patients. At the follow-up after 7.6–41.6 months (median, 26.2 months), a Fontan operation with an extracardiac conduit had been performed in seven patients at the age from 15.0 to 25.3 months (median, 22.6 month) and the others were waiting for Fontan and were in a good hemodynamic state (Fig. 3).

Table 2
Data of cardiac catheterization before BCPS (n = 9)

<table>
<thead>
<tr>
<th>Parameter</th>
<th>Mean ± S.D. (range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Qp/Qs (L/min)</td>
<td>1.4 ± 0.4 (0.7–2.3)</td>
</tr>
<tr>
<td>PAP (mmHg)</td>
<td>13 ± 4 (10–21)</td>
</tr>
<tr>
<td>PA index (cm²/m²)</td>
<td>289 ± 111 (181–454)</td>
</tr>
<tr>
<td>Rp index (unit·m⁻¹)</td>
<td>1.4 ± 0.9 (0.6–3.2)</td>
</tr>
<tr>
<td>SaO₂ (%)</td>
<td>77 ± 7 (69–88)</td>
</tr>
<tr>
<td>V-EDV (%)</td>
<td>186 ± 71 (118–309)</td>
</tr>
<tr>
<td>V-EDP (mmHg)</td>
<td>7 ± 3 (4–9)</td>
</tr>
<tr>
<td>V-EF (%)</td>
<td>63 ± 6 (57–74)</td>
</tr>
<tr>
<td>AVVR</td>
<td>None 3, trivial 5, mild 1</td>
</tr>
</tbody>
</table>

BCPS, bidirectional cavopulmonary shunt; SD, standard deviation; Qp/Qs, pulmonary to systemic flow ratio; PAP, mean pulmonary artery pressure; PA, pulmonary artery; Rp, pulmonary arterial resistance; SaO₂, arterial oxygen saturation; V-EDV, ventricular end-diastolic volume; V-EDP, ventricular end-diastolic pressure; V-EF, ventricular ejection fraction; AVVR, atrioventricular valve regurgitation.

Fig. 1. Angiography before BCPS. There is no stenosis in the pulmonary artery. The site of anastomosis is located proximal. MPA, main pulmonary artery; LSA, left subclavian artery; BCPS, bidirectional cavopulmonary shunt.

Fig. 2. Probability of BCPS achievement for all survivors. The time represents the period from mBTS to BCPS.
mBTS, modified Blalock–Taussig shunt; BCPS, bidirectional cavopulmonary shunt.

Fig. 3. Schema of postoperative course.
mBTS, modified Blalock–Taussig shunt; BCPS, bidirectional cavopulmonary shunt; PA, pulmonary artery; AV, atrioventricular valve; TAPVD, total anomalous pulmonary venous drainage; TCPC, total cavopulmonary connection.

4. Discussion

Recent improvements in the Norwood operation have demonstrated that a rather small shunt is useful in reducing excessive volume load to the ventricle and the pulmonary...
beds in a functionally single ventricle. In this study, we demonstrated the successful application of a 3-mm shunt in patients with a functionally single ventricle with duct-dependent pulmonary circulation. We successfully achieved BCPS as the second stage in the staged Fontan strategy.

4.1. Graft size and the timing of the 2nd stage

Ventricular function was maintained in a stable condition with minimum AVVR. The pulmonary artery grew well and the pulmonary vascular resistance was demonstrated to be low. We believe that a 3.0-mm BTS avoided volume overload by obliterating excessive pulmonary blood flow and thus maintained ventricular function at a reasonable level. On the other hand, even a 3-mm graft was large enough for the PA to grow adequately to attain right heart bypass. As a result, the second stage of a right heart bypass was successfully undertaken in a reasonable period in most of our patients. It appears that the minimum pulmonary blood flow using a 3.0-mm graft gave maximum gain from the vascular beds and the ventricle.

Obviously, a 3-mm BTS might not remain patent as long as a 3.5- or 4-mm shunt. Recent successful application of BCPS at an earlier age, however, could circumvent such limitations associated with a 3-mm BTS [9–13]. It has been demonstrated that the pulmonary-to-systemic flow ratio significantly increased after placement of the shunt and, therefore, the shunt provided significant volume overload to the ventricle. Therefore, we prefer earlier application of BCPS. Chang et al. [11] reported encouraging early results in terms of improved oxygenation with low morbidity and mortality in infants who underwent BCPS at an age ranging from 4.2 to 6.5 months old. Jaquiss et al. [10] revealed that early BCPS after Norwood operation did not show a significant increase in mortality compared with an older group of patients (4 months) although the rate of morbidity was higher in the younger group of patients. However, a lower age limit for BCPS does seem to exist. Reddy et al. [13] performed BCPS at an earlier age of between 0.8 and 6.0 months and recommended that BCPS should be deferred until the patient is at least two months old.

4.2. Postoperative factors

We should mention several important points for managing patient care. First, prostaglandin E1 could be changed to Prostaglandin CD at about one week before surgery so that the duct can be reduced in size several hours after surgery. This may then prevent the ventricle from acute volume overload after BTS. Second, although the duct tended to close spontaneously several days after surgery in all our patients, we do not recommend ligating a duct during surgery since a 3.0-mm shunt seemed only marginally sufficient to maintain an adequate oxygen level in the immediate postoperative period when pulmonary resistance changes. In the case that underwent sternotomy, we preferred to interpose a graft between the carotid artery and the PA to control the graft flow with adjustment of length. A significant correlation between non-confluent PA and mortality in the heterotaxy syndrome has been reported [14]. There was no PA coarctation in our series of patients, which included heterotaxy syndrome in four patients. We paid careful attention to the possibility of pulmonary coarctation during these procedures as a matter of course.

4.3. Anticoagulation therapy

Anticoagulation therapy needs to be discussed. We used heparin infusion for five days after BTS and gave oral aspirin to all patients when starting oral feeding, and warfarin in some cases. Aspirin reduces the risks of morbidity and mortality from aorto-pulmonary artery shunts [7]. Warfarin therapy is problematic because it may be hard to correctly manage coagulation level in a small baby. Although the grafts were occluded in two patients, those might be exceptional, as one had undiagnosed coagulopathy and another had long-standing high airway pressure owing to bronchomalacia.

5. Conclusion

In creating the aortopulmonary shunt, we used the smallest BTS currently available, a 3-mm shunt, and assessed the clinical outcomes. We conclude that a 3-mm BTS successfully prevented the functionally single ventricle from volume overload, allowed the growth of pulmonary vascular beds and enabled the subsequent right heart bypass operation.

References


