A Fontan completion through stage I bilateral pulmonary artery banding for hypoplastic left heart syndrome

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Although the stage I Norwood procedure is usually the first choice for hypoplastic left heart syndrome (HLHS), a neonatal Norwood operation requires complicated postoperative management, such as delayed sternal closure or prolonged mechanical ventilation, and interstage deaths 1 after stage I Norwood procedures are an issue. Here we report less invasive Fontan completion through a novel strategy consisting of bilateral pulmonary artery (PA) banding for stage I palliation, followed by the Norwood procedure concomitantly performed with bidirectional Glenn shunt (BDG) for stage II palliation for HLHS.

Clinical Summary

A 7-day-old boy weighing 3.5 kg had a diagnosis of HLHS with aortic stenosis and mitral stenosis associated with mild tricuspid regurgitation (TR) without noncardiac anomalies. Immediately after admission, prostaglandin E₁ (PGE₁) was administered. Atrial septal defect (ASD) was not restrictive, and mechanical ventilation was not required before the operation. For stage I palliation, we performed bilateral PA banding through a median sternotomy with a right circumference of 10 mm and a left circumference of 10.5 mm with a handmade banding tape in which an expanded polytetrafluoroethylene (ePTFE) sheet of 0.4 mm thickness was trimmed to 2 mm width. The sternum was closed in the operating room, and extubation was performed on the operative day, with inotropic support finishing the next day. PGE₁ administration (5 ng/[kg · min]) was continued with intensive care unit hospitalization until stage II palliation. We performed balloon atrial septostomy when the patient was 2 months old because of progressive restrictive ASD. The mean right PA pressure was reduced to 16 mm Hg after balloon atrial septostomy, bilateral PA was balanced, and Nakata index was 168 mm²/m² on angiocardiology (Figure 1, A). The ductus arteriosus was kept patent, and there was no transductal pressure gradient before stage II (Figure 1, B). Although TR had progressed to moderate according to echocardiog-

raphy, mechanical ventilation was not required, and neither necrotizing enterocolitis from enteral feeding or infection from PGE₁ infusion occurred in the interval to stage II.

When the patient was 3 months old and weighed 4.5 kg, we performed a Norwood procedure that consisted of coronary flow and aortic arch reconstruction and ASD enlargement, BDG, and tricuspid annuloplasty for TR. Procedures were performed through a median sternotomy with cardiopulmonary bypass without circulatory arrest.² PA plasty was not required on the banding site, only PA debanding. The sternum was closed in the operating room. Inotropic support was finished on the operative day, and mechanical ventilation was continued for 2 days. After stage II palliation, there was no recoarctation of the aorta or restrictive ASD, and TR was trivial. Nakata index was 171 mm²/m² without PA distortion, and PA mean pressure was 15 mm Hg before the Fontan procedure.

When the patient was 2 years 4 months old and weighed 12.0 kg, we performed an extracardiac total cavopulmonary connection with an 18-mm ePTFE conduit with fenestration by a 5-mm ePTFE conduit. The patient was extubated on the operative day, and chest drainage was continued for 3 days. Postoperative angiocardiology showed adequate growth of the PA without distortion (Figure 2), and mean PA pressure was 9 mm Hg. The patient is alive and well after an 8-month follow-up.

Discussion

We report an easier postoperative management of staged Fontan completion through stage I bilateral PA banding for HLHS. A low mortality has been reported after Fontan completion in HLHS³; thus staged palliative management is important. The merits of this strategy are as follows: (1) Bilateral PA banding for HLHS is less invasive. (2) The reduction of interstage mortality before stage II palliation can be expected by maintaining hospitalization for PGE₁ administration. (3) The Norwood procedure concomitantly performed with BDG reduces volume overload and promotes hemodynamic stability. For this strategy, the most important issues are restrictive ASD⁴ and maintenance of systemic flow. Advanced catheter techniques would be required for certain ASD openings. For maintenance of systemic flow, although ductal stenting has been reported,⁵ we think PGE₁ administration naturally forms a patent ductus arteriosus without any complications of injury and iatrogenic narrowing of a weak ductus arteriosus. In our experience of this strategy for 6 patients with HLHS managed by PGE₁ administration, we found there was a longer period, 4 months, of ductal opening without narrowing. We hope later to report on the molecular mechanisms of ductal opening.

In conclusion, less invasive Fontan completion is possible through bilateral PA banding for stage I palliation, followed by the Norwood procedure concomitantly performed with BDG for stage II palliation. Progressive management for restrictive ASD and the

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Figure 1. Angiocardiography at 2 months. A, Arrows indicate bilateral PA banding sites. B, Arrows indicate patent ductus arteriosus.

Figure 2. Angiocardiography after Fontan completion.

maintenance of systemic flow ensures the integrity of this bilateral PA banding strategy for patients with HLHS.¹

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